DIAGNOSIS OF CONGENITAL HEART DISEASE

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DIAGNOSIS

A Clinical and Technical Study by

OF CONGENITAL HEART DISEASE

the Cardiologic Team of the Pediatric Clinic
Karolinska Sjukhuset, Stockholm

SECOND EDITION

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Preface to Second Edition

THIS SECOND EDITION is based on clinical and roentgenologic studies of 742 cases of congenital heart disease investigated between October 1951 and July 1957. Since publication of the first edition, the case material has been almost doubled, and new types of congenital heart disease have been observed. By application of new methods of examination and modification of the earlier ones, it has been possible to make further differentiations in certain types of heart disease, which have led to some corrections in the descriptions given in the first edition. This applies in particular to atrial septal defects and anomalous drainage of pulmonary veins. Four completely new chapters have been added.

In addition, an account is given of the effect of exercise on the hemodynamics in a small number of adults with congenital heart disease These patients are not otherwise included in the case material The investigations were made at the Department of Clinical Physiology (Director: Professor Torgny Sjostrand), Karolinska Sjukhuset.

During the past few years, great advances have been made in cardiology; these have been reflected in an exceedingly ex- [illegible] fully aware that some important works may be lacking in our list of references. This lack is to be ascribed to the fact that this book is intended as an account of our own observations and not as a comprehensive survey of the literature.

We wish to express our sincere gratitude to all those who have helped us in preparing this second edition. Our particular thanks are due to Drs. Danae Ikkos, Bengt Strindberg, Mats Barr, Carl Ove Ovenfors, and Erik Carlsson, as well as to Mrs. Margaretha Ibring, Miss Gun Persson, Mr. Borje Nilsson, who prepared the photographs, and Mrs. Erica Odelberg, who translated the manuscript from the Swedish.

—THE AUTHORS

Stockholm
April, 1958

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An account is given of the results obtained with the diagnostic methods used, with particular emphasis on the special technical aids which make possible a detailed functional and anatomic diagnosis. Evaluation of the findings at the customary clinical examinations has been greatly facilitated by these means. The reason underlying the high incidence of extensive special investigations was to promote one of the main objects of this book, namely, to appraise the value of the respective methods.

The results of the physical examinations, including phonocardiography, have been analyzed by Edgar Mannheimer and Bengt Jonsson, and those of the other clinical investigations and of cardiac catheterization by Bengt Jonsson. Sven Roland Kjellberg and Ulf Rudhe are responsible for the interpretation of the roentgenologic and angiocardiographic findings, and Ulf Rudhe for the observations at electrokymography.

Space does not permit us to thank all those who have helped in the preparation of this book. We nevertheless wish to express our particular gratitude to the following:

To Arvid Wallgren, for his encouragement during the course of our work.

To Leo G Rigler, Jack Friedman and Fred Jenner Hodges, who have shown great interest in our project.

To Clarence Crafoord, Gunnar Ekstrom, Viking Olof Björk and Ake Senning, for many stimulating discussions.

To Lennart Rahm, for valuable theoretical views on hemodynamics

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1 Embryologic Survey

THIS CHAPTER has no pretensions to being a complete account of the whole course of development of the circulatory system. Its object is merely to elucidate, from the embryologic viewpoint, the malformations of the heart and great vessels that comprise the clinical material in our book. The data are based mainly on current conceptions of the process of development (18, 101, 102, 125, 181, 217, 261, 526, 527, 567, 576, 646, 647) (See also the anatomic background in the relevant chapters)

A. DEVELOPMENT OF THE GREAT VEINS

1. The Systemic Veins

The circulatory system is laid down at a very early stage of fetal development. According to Eternod (243), a closed blood circuit already exists in a human fetus about 1 3 mm long, at the end of the second embryonic week. This circuit consists of the heart, two aortae, which pass caudally into the umbilical arteries, and two *umbilical veins*. The task of these veins is to carry the blood from the capillaries of the chorion directly to the heart. Further development takes place rapidly, and in a subsequent stage branches grow out from the umbilical veins and unite with the newly formed vessels in the yolk sac (526, 527) These venous branches increase rapidly in size and form the so-called *omphalomesenteric* veins, which open into the umbilical veins slightly caudal to the heart. Because of the increased blood flow through the parts of the umbilical veins lying cranially to the site of entry of the omphalomesenteric veins, they become widened into a *sinus venosus* (Fig 1).

The blood which circulates within the embryo is, on the contrary, collected by the so-called *cardinal veins*. These are laid down in the third embryonic week (244) as two paired venous trunks. One pair, the *anterior cardinal veins*, arises from the cranial part of the body; the other pair, the *posterior cardinal veins*, arises from the caudal part. These cardinal veins unite on either side of the heart into a short main trunk, the *common cardinal veins* or *ducts of Cuvier* They in turn [illegible]

is because these veins drain not only the segmental veins of the thoracic, abdominal and pelvic regions, but also the veins from both the lower and the upper extremities. As the heart descends caudally, the orifices of the *subclavian veins* are, however, displaced cranially, so that they will gradually open into the *anterior cardinal veins*.

After this occurrence, a wide anastomosis forms between the anterior cardinal veins. the *left innominate vein* (Fig 2, D–F). The part of the right cardinal vein which lies caudal to the site of anastomosis

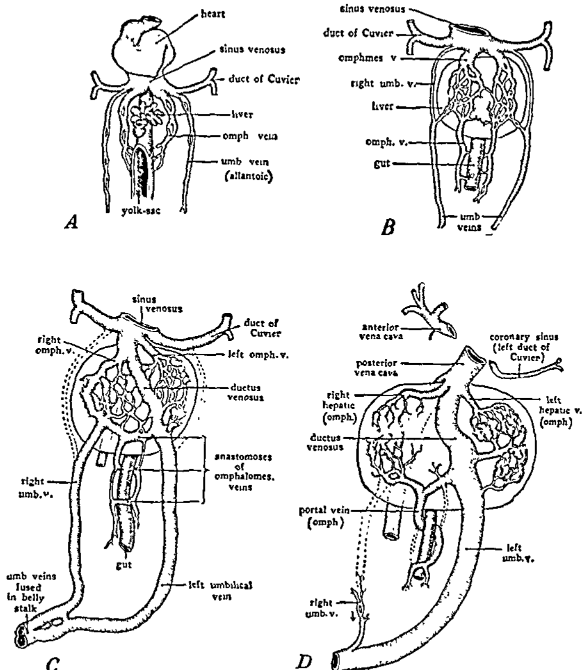
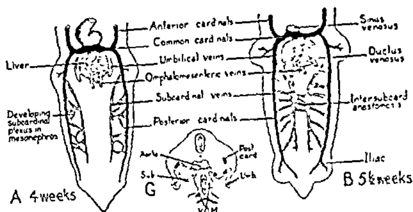
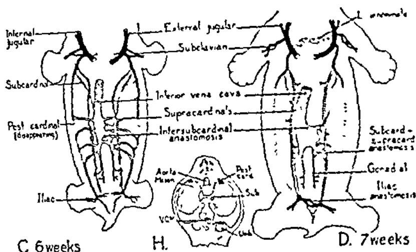
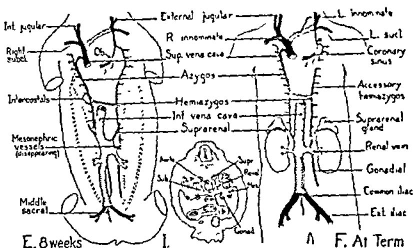


Fig. 1.—Diagrams showing development of hepatic portal circulation from omphalomesenteric veins, and changes by which blood returning from placenta by way of umbilical veins is rerouted through the liver. *A*, based on conditions in pig embryos of 3–4 mm, applicable to human embryos of fourth week; *B*, based on pig embryos of about 6 mm, applicable to human embryos


Anterior cardinals
Common cardinals
Umbilical veins
Omphalomesenteric veins
Subcardinal veins
Posterior cardinals
Liver
Developing subcardinal plexus in mesonephros
Sinus venosus
Ductus venosus
Intersubcard anastomosis
Iliac
Aorta
Post card
Sub
Umb
VOM
A 4weeks
G
B 5½weeks


Internal jugular
External jugular
Subclavian
L innominate
Subcardinal
Inferior vena cava
Supracardinals
Post cardinal (disappearing)
Intersubcardinal anastomosis
Subcard-supracard anastomosis
Gonadal
Iliac
Iliac anastomosis
Aorta
Post card
Sub
Umb
C 6weeks
H.
D. 7weeks


Int. jugular
External jugular
L. Innominate
R innominate
L. subcl
Right subcl.
Sup. vena cava
Coronary sinus
Azygos
Accessory hemiazygos
Intercostals
Hemiazygos
Inf vena cava
Suprarenal gland
Suprarenal
Renal vein
Mesonephric vessels (disappearing)
Gonadial
Common iliac
Middle sacral
Ext. iliac
Aorta
Sub
Supr
Renal
Gonad
E. 8weeks
I.
F. At Term

forms, together with the common cardinal vein, the primordium of the *superior vena cava*. The part lying between the site of anastomosis and the right subclavian vein becomes the *right innominate vein*. At this stage, a *left superior vena cava* also exists. It is formed by the left cardinal vein, below the origin of the left innominate vein, and (664). The blood from the left upper part of the body then continues to flow downward through a *persisting left superior vena cava*, as shown in Figure 476 (p. 527). Even when an anastomosis is present, there may be *double venae cavae* (Fig. 5).

The fate of the *posterior cardinal veins* is so intimately associated with the develop-

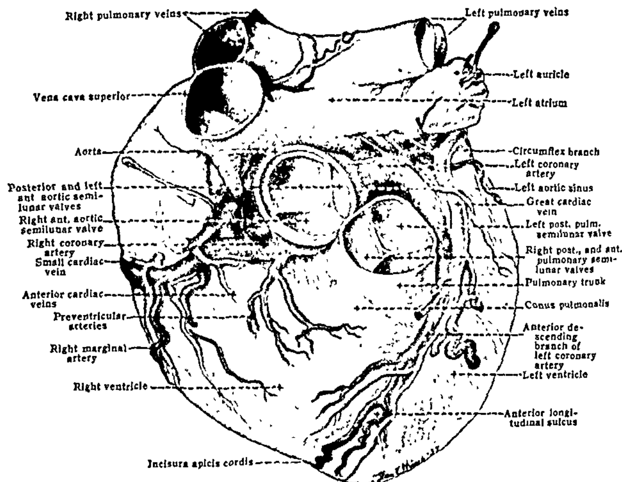


Fig. 3a.—Cephalic view of the heart with the epicardium removed to expose the injected coronary vessels. (From *Morris' Human Anatomy* [10th ed., New York Blakiston Company, 1942]) (*Continued.*)

the left common cardinal vein. The greater part of this caval vein soon atrophies (268). Only the caudal part persists, in the form of the *oblique vein of the left atrium* (Fig. 3), as well as the cranial part, in the form of the *first intercostal vein* (Fig. 4). We shall return to this matter on page 12.

Considerable deviations frequently occur. Sometimes no anastomosis takes place between the two anterior cardinal veins

ment of the *hepatic veins* and the *inferior vena cava* that a short account of this process is necessary.

The two omphalomesenteric veins run through the mesenchymal septum transversum, which separates the caudal part of the pericardial cavity from the pleural and abdominal cavities (102, 460) In connection with the development of the medial lobe of the liver, two vascular networks are

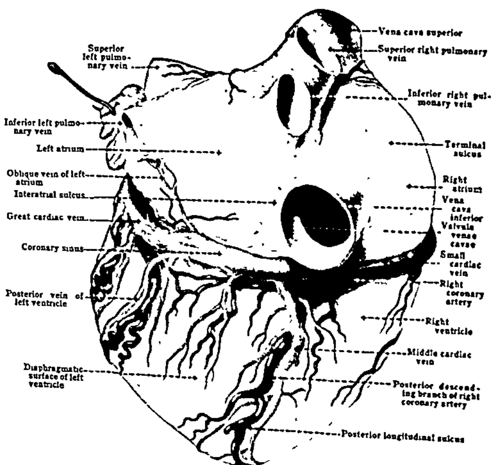


Fig. 3a (*cont.*) —Dorsocaudal view of the heart.

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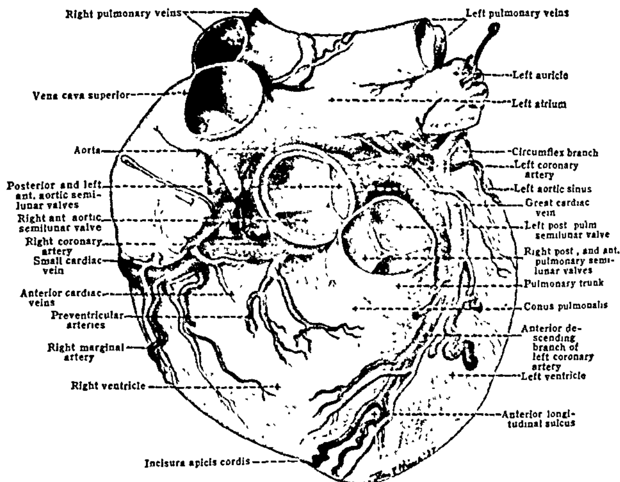
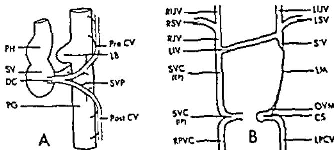


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ugular veins, *RJV* and
postcardinal veins, *RSV*
SVC (EP) and SVC (IP),
and Oram, Brit Heart J. 15:305, 1953.)

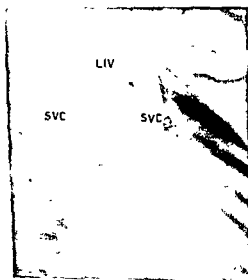


Fig 5 —Left (*SVCs*) and right (*SVC*) superior vena cava. The left innominate vein (*LIV*) is narrow (Courtesy of Dr F Ulfsparre, Stockholm.)



Fig. 3b—Persistent truncus arteriosus Girl, aged 17 (S A 361223) At the end of the injection, the catheter slips out into the right atrium and the coronary sinus As seen also in Figure 308 (p 334), the left superior vena cava is not entirely obliterated, a small lumen persists, extending almost up to the left innominate vein *AO*, aorta, *LAC*, left anterior cardinal vein, *LPA*, left branch of pulmonary artery, *Tr*, truncus

hens than through the left. According to Broman (101, 102), this accounts for the *left vena revehens* becoming successively narrower and finally disappearing altogether at the beginning of the fourth embryonic week. If this were the case, it could be expected that, in a rotation anomaly of the heart in which the right atrium lies to the left of the midline, the left vena revehens would persist and the right would

tal vein. When this communication is e[...] tablished, the part of the *left umbilical ve[...]* above the anastomosis atrophies, and [...] blood then flows entirely through the port[...] system of veins into the sinus venosu[...] Because of the successively increasi[...] blood flow through the rapidly growing pl[...] centa and the wide, persisting left umbi[...] cal vein, a continuous dilatation of th[...] intrahepatic vessels takes place. This a[...]



Fig. 7.—Tetralogy of Fallot [...] (P.K. 491010) [...] ance [...] men, [...] vena [...]

atrophy. This may possibly apply in the case illustrated in Figure 7.

Owing to the increased blood flow through the *right vena revehens*, this ves-[...]

[...] revehens atrophies, changes occur in the *umbilical veins* running close to the liver. At the end of the third embryonic week, these veins still run as two equally thick vessels up to the sinus venosus (Fig. 1). However, the *right umbilical vein* soon starts to atrophy, and the left joins the most cranial transverse anastomosis of the por-

plies in particular to those *offering* th[...] shortest route between the umbilical vei[...] and the *common* vena revehens. In thi[...] way, the *ductus venosus arantii* arises (Fig[...] 6).

During the initial developmental stages of the fetus, the *posterior cardinal vein[...]* [...]

[...] the *subcardinal veins* (Fig. 2). As their growth progresses, they collect an increasing amount of blood from the mesonephric region and partly take over the function of the cardinal veins. When these subcardinal veins first appear, they

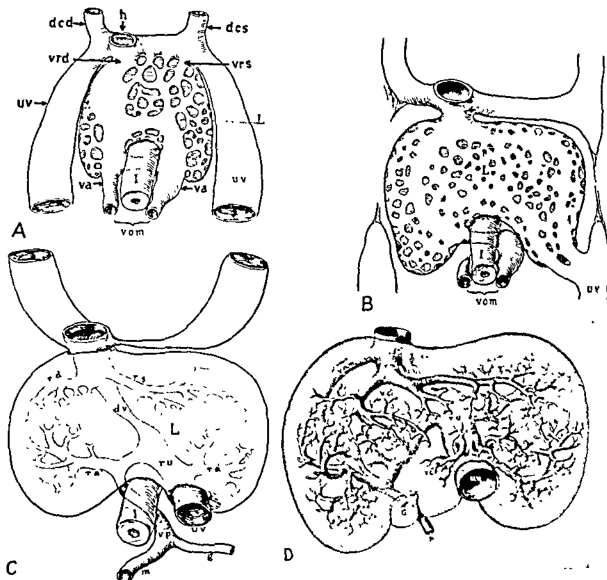


Fig. 6.—Semidiagrammatic reconstruction of veins of the liver of a human embryo. *A*, 4.5 mm embryo, *B*, 4 mm, *C*, 9 mm, *D*, 11 mm, *dcd* and *dcs*, right and left ducts of Cuvier, *dv*, ductus venosus aranthi, *g*, gastric vein, *h*, opening of sinus venosus into heart, *I*, intestine, *L*, liver, *m*, mesenteric vein, *ra*, ramus arcuatus, *rd*, ramus dextra, *rs*, ramus sinistra, *uv*, umbilical vein, *va*, venae advehentes, *vom*, omphalomesenteric vein, *vrd* and *vrs*, right and left venae revehentes (From Mall, Am. J Anat 5:227, 1906.)

formed from the parts of the omphalomesenteric veins running in the septum transversum. These two networks nevertheless soon fuse, through the development of anastomoses, into *one* vascular network with two afferent vessels, the *venae advehentes*, and two efferent vessels, the *venae revehentes hepatis*. Through the occurrence of three powerful anastomoses, as well as atrophy of certain parts of the venae advehentes and revehentes (Figs. 1 and 6), a single vena advehens hepatis is formed. It surrounds the gut in a spiral and opens into the right half of the liver (339). This vein then joins the *mesenteric vein* and the *splenic vein* to form the *portal vein* (340)

As early as the third embryonic week, the opening of the sinus venosus into the heart begins to be shifted to the right of the midline As a result, the blood probably flows more easily through the right vena reve-

hens than through the left. According to Broman (101, 102), this accounts for the *left vena revehens* becoming successively narrower and finally disappearing altogether at the beginning of the fourth embryonic week If this were the case, it could be expected that, in a rotation anomaly of the heart in which the right atrium lies to the left of the midline, the left vena revehens would persist and the right would

blood then flows entirely through the portal system of veins into the sinus venosus. Because of the successively increasing blood flow through the rapidly growing placenta and the wide, persisting left umbilical vein, a continuous dilatation of the intrahepatic vessels takes place. This ap-



Fig 7.—Tetralogy of Fallot with infundibular stenosis and rotation of heart. Boy, aged 4 (P K. 491010), see Figure 253 (p. 272). Abdominal viscera are not transposed, despite appearance in *B* The right atrium (*RA*) lies to the left. The inferior vena cava, to the right in the abdomen, deviates to the left and crosses the spine before opening into the right atrium *VRS*, inferior vena cava (+ left vena revehens).

atrophy. This may possibly apply in the case illustrated in Figure 7

Owing to the increased blood flow through the *right vena revehens*, this vessel becomes increasingly dilated and, after complete atrophy of the left vena revehens, becomes known as the *common vena revehens* At about the same time that the left vena revehens atrophies, changes occur in the *umbilical veins* running close to the liver At the end of the third embryonic week, these veins still run as two equally thick vessels up to the sinus venosus (Fig 1). However, the *right umbilical vein* soon starts to atrophy, and the left joins the most cranial transverse anastomosis of the por-

plies in particular to those offering the shortest route between the umbilical vein and the common vena revehens. In this way, the *ductus venosus arantii* arises (Fig. 6).

During the initial developmental stages of the fetus, the *posterior cardinal veins* are the only veins draining the caudal part of the body. In the sixth fetal week, a group of new vessels starts to appear in the mesonephric region: the *subcardinal veins* (Fig. 2). As their growth progresses, they collect an increasing amount of blood from the mesonephric region and partly take over the function of the cardinal veins. When these subcardinal veins first appear, they

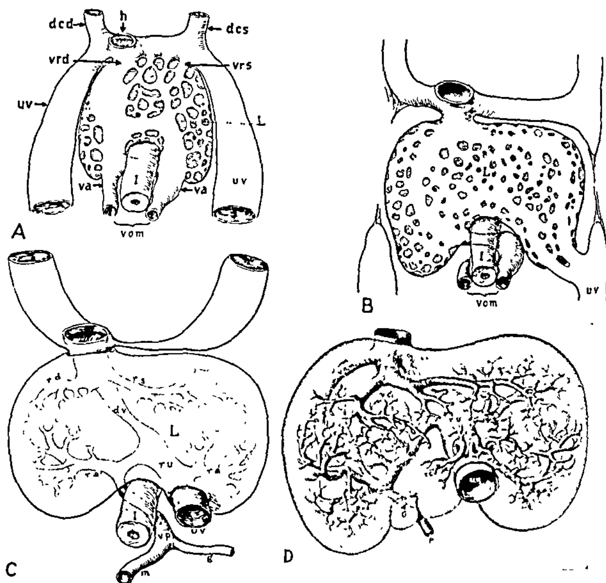


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form an irregular plexus which empties directly into the posterior cardinals. Longitudinal channels are, however, formed in the ventromedial border of the mesonephric ridge and gradually dilate into the *main subcardinal veins* They run almost parallel to the posterior cardinal veins and empty into their cranial portion (Fig 2, C).

With the growth of the mesonephric ridge, the rapidly growing subcardinal veins become shifted increasingly close to out in the caudal direction from the right vena revehens as early as the fourth embryonic week. According to McClure *et al.* (449, 450) and Reagan (549), it is, instead, the capillaries which branch into the mesentery, between the liver and the mesonephric ridge, that convey this blood flow. However this may be, these vessel paths become widened and take over an increasing amount of the function from the posterior cardinal veins. This newly formed

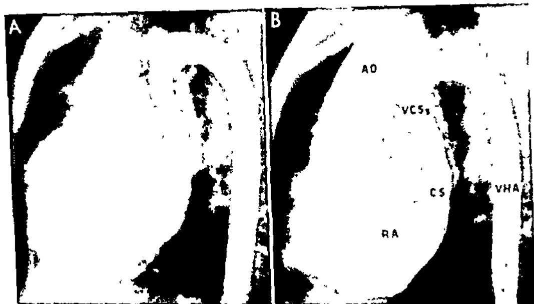


Fig 9.—Absence of superior part of the inferior vena cava Boy, aged 9 (A M. 450921). The lower part of the body is drained chiefly through the hemiazygos vein (*VHA*), which empties into the left superior vena cava (*VCSs*) *AO*, aorta, *CS*, coronary sinus, *RA*, right atrium.

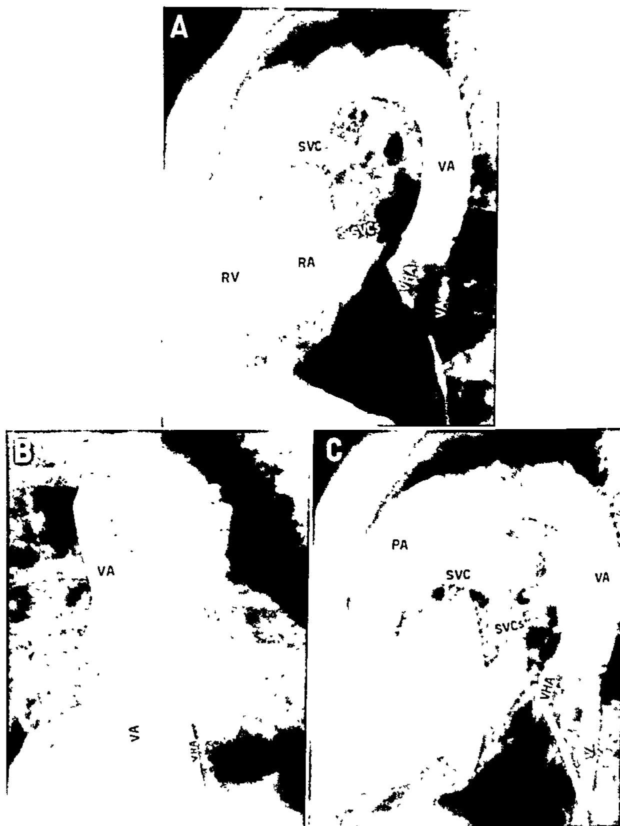
each other. Small anastomoses are gradually established between them, they soon become dilated and finally form a wide *intersubcardinal anastomosis* (Fig 2, C and D) The *subcardinal venous sinus* then formed probably offers considerably less resistance to the blood than do the cardinal veins According to Butler (124), this is presumably the reason why the blood follows this path, and the cardinal veins in the mid-mesonephric region atrophy

Wit[illegible]

ver[illegible]

cat[illegible] the heart In the opinion of Broman (102), a small vein, which meets the subcardinal veins, grows channel is known as the *primitive inferior vena cava* (Fig 2, C–F).

The result of this rerouting of the blood flow is that the cardinal veins atrophy, with the exception of the most cranial portion, above the site of entry of the subcardinal veins In addition, the subcardinal veins are reduced to two narrow, longitudinal vessels, the right corresponds to the *azygos vein* and the left to the *hemiazygos vein* (Fig. 2, E–F). At the same time, an anastomosis is established between these veins, in the cranial part of their course Probably on the grounds of the blood then being shunted, via this anastomosis, from the left to the right subcardinal vein, the portion of



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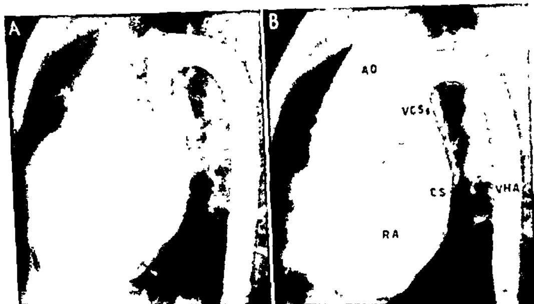


Fig. 9.—Absence of superior part of the inferior vena cava. Boy, aged 9 (A.M. 450921). The lower part of the body is drained chiefly through the hemiazygos vein (*VHA*), which empties into the left superior vena cava (*VCSs*). *AO*, aorta, *CS*, coronary sinus, *RA*, right atrium.

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With the development of the subcardinal venous sinus, a new more direct communication arises between it and the heart. In the opinion of Broman (102), a small vein, which meets the subcardinal veins, grows channel is known as the *primitive inferior vena cava* (Fig. 2, C–F).

The result of this rerouting of the blood flow is that the cardinal veins atrophy, with the exception of the most cranial portion, above the site of entry of the subcardinal veins. In addition, the subcardinal veins are reduced to two narrow, longitudinal vessels, the right corresponds to the *azygos vein* and the left to the *hemiazygos vein* (Fig. 2, E–F). At the same time, an anastomosis is established between these veins, in the cranial part of their course. Probably on the grounds of the blood then being shunted, via this anastomosis, from the left to the right subcardinal vein, the portion of

the left subcardinal vein cranial to the anastomosis atrophies wholly or partly. It sometimes persists as a narrow vessel, the *accessory hemiazygos* (Fig. 2, *F*). If anastomosis does not take place, or it is incomplete, the hemiazygos vein subsequently empties all or part of its blood into the persisting superior segment of the left cardinal vein. Figure 478 (p. 529) shows such a case, in which the hemiazygos vein empties *into the left innominate vein via the left anterior cardinal. It may be inferred from this figure that the azygos vein cannot be identified roentgenologically and that at any rate* its superior segment is probably lacking.

This complicated interplay between new formation and atrophy would seem to give ample scope for various kinds of malformations. Notwithstanding, vascular anomalies of any major importance in this region were relatively uncommon in our series. In addition to the case just mentioned, we observed a malformation of this kind in three *other cases (e.g., Fig. 475, p. 526). It is seen* that the azygos vein is greatly dilated, because it drains almost all of the lower part of the body. Presumably, the new formation of vessels between the subcardinal venous sinus and the liver had never taken place, and the prerequisite for the formation of the superior segment of the inferior vena cava was therefore lacking (410, 625). Consequently, the blood from the *subcardinal venous sinus must continue to* flow through the subcardinal and cardinal veins (the azygos, Figure 8, and hemiazygos, Figure 9) As may be inferred from Figure 8, the junction of the azygos and the hemiazygos is at a considerably more caudal level than normally (normally, it is at the level of the sixth to seventh thoracic vertebrae). Moreover, the inferior vena cava is lacking, and the blood from the *lower part of the body is therefore drained* chiefly into the superior vena cava via the azygos vein.

2. The Sinus Region

The *sinus venosus* originally denotes the junction of the great veins which return *the blood to the heart. It nevertheless* undergoes such radical changes in shape and position during the course of development of the heart that its original appearance is entirely lost. In a 4 to 5 week embryo, a partial condensation has already taken place, by fusion of the cephalic parts of the omphalomesenteric veins (Fig. 10). The sinus venosus then forms a partly transverse vessel, which continues on either side into a *limb directed backward and upward*, the so called *horn of the sinus venosus* (526). The common cardinal vein of the corresponding side opens in the tip of this horn (Fig. 10). Owing to the expansion of the atrium, the rotation of the heart and the shift of the opening of the sinus to the right in the atrium, the sinus venosus forms, together with the cardinal veins, a figure reminiscent of a U.

The increasing growth of the inferior vena cava, as well as the transformation of the right anterior and common cardinal *veins into the superior vena cava*, result in rapid changes in the conditions in the sinus region. The blood from the left anterior cardinal vein is shunted through the left innominate vein to an increasing extent to the right side. This, in addition to the disappearance of the left vena revehens and *the proximal part of the left umbilical vein*, causes a decrease in the blood flow through the left anterior cardinal, the left common cardinal vein and the *left horn* of the sinus, and a reduction in their width (Fig 10, *E*). From this stage onward, the rate of the aforementioned vessels differs. The left anterior cardinal vein atrophies almost entirely caudal to the origin of the left innominate vein, but may persist as a narrow, *hollow strand of varying length* (Fig 3). The remains of the cardinal vein, which crosses the dorsal wall of the left atrium, is usually known as the *oblique vein of the left atrium* or the *vein of Marshall* (Figs. 3 and 10, *F*). In about the seventh to eighth embryonic week, small vessel branches are formed from *both the heart and the left* common cardinal vein, they assemble the blood from the heart itself (Fig. 10, *E*) Thereafter, the left common cardinal vein

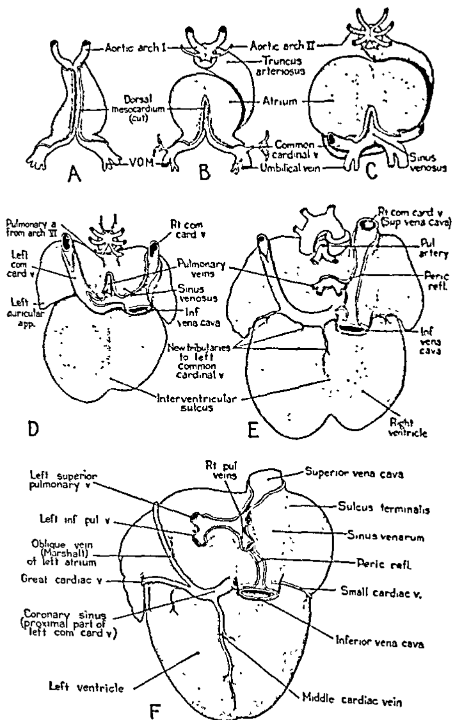


Fig 10 —Six stages in development of the heart, drawn in dorsal aspect to show changing relations of sinus venosus and great veins entering heart. *A*, embryo at 2½ weeks (8–10 somites), *B*, 3 weeks (12–14 somites), *C*, 3½ weeks (17–19 somites), *D*, 5 weeks (6–8 mm C-R); *E*, 8 weeks (embryos about 25 mm), *F*, 11 weeks (embryos about 60 mm). (From Patten, B. M.: *Human Embryology* [New York Blakiston Company, 1946])

and the left horn of the sinus venosus partake in the formation of the *coronary sinus* (Fig. 10, *F*). This may (90, 452, 483, 549) in rare cases open into both the right and the left atrium (Fig. 446, p. 488) or into the left atrium alone (44, 358, 483, 520) or communicate only with the left superior vena cava (419, 647).

In contrast to the left horn of the sinus venosus, the *right horn* increases successively in size, owing to the continuous increase in the blood flow through the superior and inferior venae cavae. In the adult heart, this horn forms the *sinus venarum*.

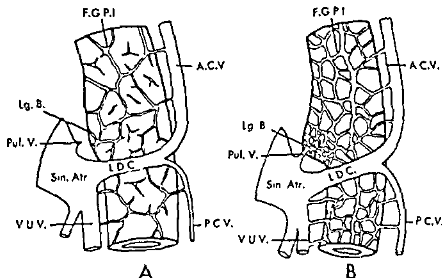


Fig. 11.—Diagrams illustrating formation and routes of drainage of the foregut plexus in the rat *A*, 3 mm rat embryo, stage 1. The foregut plexus is not yet complete and is draining into the cardinal and vitelloumbilical veins. The cardiac part of the pulmonary vein is seen as an endothelial outgrowth from the dorsal part of the sinoatrial chamber and has not yet "tapped" the fore-

3. The Pulmonary Veins

Concurrently with the growth, during the third to fourth embryonic weeks, of the pulmonary primordium, a *primary venous plexus* is developed, which drains the area for the lung buds (Fig. 11). It empties through a group of wider channels into the anterior cardinal veins (268) During subsequent embryonic development, a direct communication is, however, formed between this venous plexus and the left atrium. A new vessel, the *common pulmonary vein*, collecting blood by a tributary vein from each lung, is laid down in the midline (cf. Fig. 10, *E* and *F*). His (338) demonstrates the pulmonary vein as opening into the left part of the sinus venosus in an early stage in the human embryo The opening is situated to the left of the common opening of the anterior and posterior cardinals (249, 580) and to the left of the valve of the sinus (106, 182, 338). The shift in position of the opening to its definitive site results from progression of this valve to the left to join the atrial septum. As early as the first half of the second embryonic month, this new venous trunk starts to undergo changes The proximal part becomes widened into a funnel shape and is incorporated with the atrial wall. The dilatation continues successively far peripherally, until the entire vein has become fused with the atrial wall The two tributary veins, the *pulmonary veins*, there-

ly open directly into the left atrium. These veins undergo the same funnel-shaped dilatation as far as the first main bifurcation on both sides. They thus take part in the formation of the dorsal wall of the atrium (106, 182). According to Keith (378), it is highly probable that parts of the sinus venosus also contribute to formation of this wall (cf. p. 54, ontogenesis). When the process is complete, all four pulmonary veins open directly into the left atrium.

The course of development just described throws light on most of the developmental anomalies found in this vascular region. If aforementioned pulmonary vein, the drainage takes place, in the majority of cases, through the left anterior cardinal vein (see also Chapter 15). An explanation is provided by the embryologic process. This cardinal vein empties either via the left innominate vein into the right anterior cardinal vein—superior vena cava—(Figs. 457 and 459, pp. 508 and 510) or the coronary sinus (Fig. 453, p. 500; Fig. 460, p. 511). In those cases in which communications have been found to the right pulmonary lobes only, or to single lobes on the left side, these veins have opened into

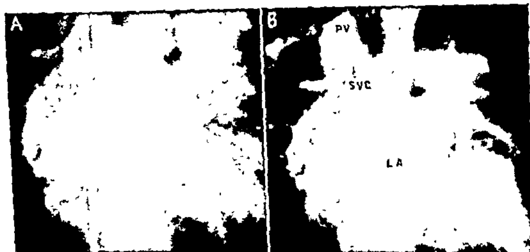


Fig 12.—Incomplete anomalous venous return Girl, aged 15 (E.I. 410918). The superior pulmonary vein on the right side (*PV*) opens into the superior vena cava (*SVC*). The volume of the left atrium (*LA*) is therefore reduced, and its upper right part is lacking

the aforementioned common pulmonary vein does not develop at all, or develops abnormally, the blood from those parts of the venous plexus that are then not drained by this vein continues to empty into the cardinal veins, usually the anterior cardinals. Because of the increased blood flow arising in this way, there is dilatation of the cardinal vein or veins involved. If such a communication with the left anterior cardinal vein persists, the normally occurring atrophy of this vein does not appear to take place Our material indicates that if no communication arises between the total plexus or its superior part—probably corresponding to the upper lobes—and the the right anterior cardinal vein (Fig. 472, p. 523), the left subclavian artery (508, 547, 687), the vena porta (517), or the right atrium. Exceptions are, however, found (116, 222, 224, 529.)

It has not yet been elucidated whether the opening of one or several pulmonary veins directly into the right atrium is due to an abnormal position of the atrial septum or to anomalous formation of the pulmonary veins. Our observations indicate that these abnormal veins generally open into the sinus venarum or its vicinity. In our opinion, this gives reason to suspect that these pulmonary veins may be remains of the channels which drain the primary

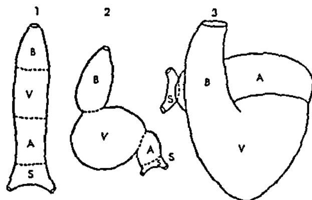


Fig. 13.—Early stages of development of the heart. *A*, atrium; *B*, bulbus cordis (conus); *S*, sinus venosus; *V*, ventricle. (From Pichon, Nouveau traité de méd. *10:2,601, 1933.*)

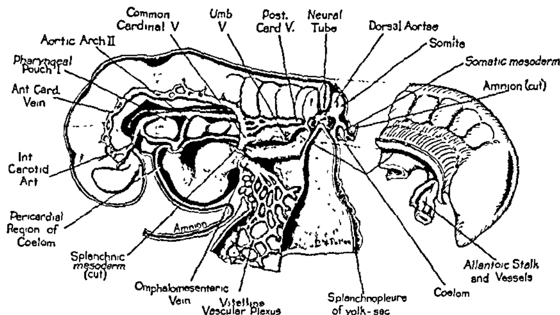


Fig. 14.—Schematized lateral dissection of human embryo of about 3 weeks to show continuity of pericardial portion of coelom with paired coelomic chambers of midbody region. (From Patten, B. M.: *Human Embryology* [New York. Blakiston Company, 1946])

venous plexus and that they have been drawn into the right atrium in connection with the incorporation of the sinus venosus with the atrium.

The dorsal wall of the left atrium is formed for the greater part by the incorporated pulmonary veins The volume of the atrium decreases if one or several of these veins does not achieve communication with the atrium, but empties elsewhere (Fig. 12 and Fig. 80, p. 78). In the former of these cases, the right superior pulmonary vein empties into the right anterior cardinal vein. In the latter case, all the lobes of the lung are drained through the left anterior cardinal vein. Similar observations have been made by Edwards *et al.* (224), Didion (191), Stoeber (630), and others.

B. DEVELOPMENT OF THE HEART

From two small groups of mesoblasts in the caudal part of the head-fold which at the end of the second embryonic week fuse in the midline into an elongated unpaired tube, a four-chambered organ, provided

Fig 15.—Ventral views of human embryonic hearts to show bending of cardiac tube and establishing of regional divisions (From Kramer, Am. J. Anat. 71:313, 1942)

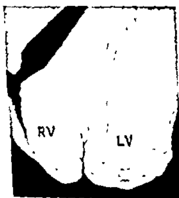


Fig. 16.—Notch at apex between left and right ventricles.

with valves, is developed in a relatively short space of time. It is capable of maintaining the circulation through two separate systems (101, 102, 181, 337, 526, 646).

The *truncus arteriosus* is given off from the cranial part of the aforementioned tube, and the great veins empty into its caudal part (*Fig. 13*). *A factor which* greatly contributes to the transformation of this tube *atrium* and the *sinus venosus* (Fig. 13). During the formation of the cardiac loop, these *grooves* become deeper, and at the same time a new groove appears in the medial segment of the *apical part* of the ventricular loop (*Fig. 15*). *This groove, which* can sometimes be seen distinctly in the fully developed heart (Fig. 16), indicates the first division of the heart into a left and a right side. There is a concurrent marked

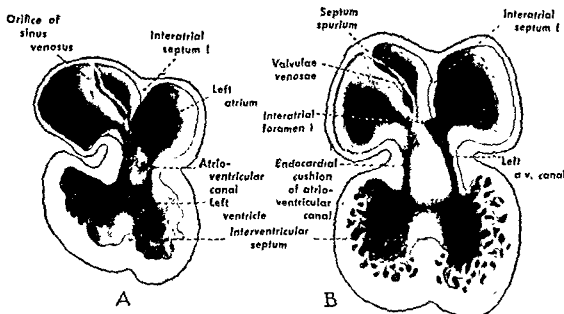


Fig. 17.—Semischematic drawings of inferior aspect of heart to show initial steps in its parti-

is its rapid increase in length. Owing to lack of space, it is at first bent sideways, but subsequently curves in a pronounced loop (Figs. 14 and 15). Since the cephalic part of the heart is stabilized by the truncus arteriosus and the caudal part by the great veins, the middle part undergoes the greatest changes in position

Even at an early stage, small grooves appear on the external aspect of the heart tube, marking its four different sections the *conus (bulbus cordis)*, the *ventricle*, the increase in the size of both the atrium and the ventricle. The atrium enlarges to such an extent that it bulges on either side of the truncus arteriosus and the conus Development then proceeds rapidly, and at the end of the first embryonic month it is so far advanced that the final division of the heart into four chambers is discernible From this stage onward, it is no longer possible to follow the development of the heart as a unit, but each of its parts must thereafter be dealt with separately.

1. The Atria and the Atrioventricular Canal

The actual division of the common atrium into two chambers takes place during the second fetal month. Complete separation does not, however, occur until after birth, when the placental circuit has been cut off and the pulmonary circulation has become stabilized. The internal separation

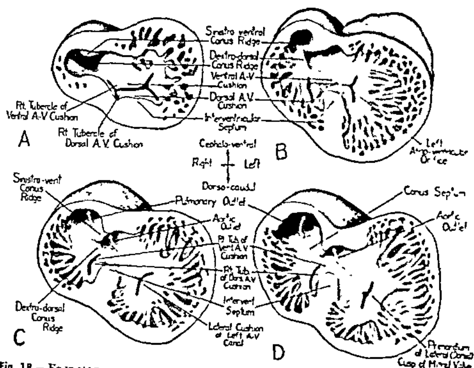


Fig 18.—F...
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of the two atria is effected by the growth of two septa, in view of the time of their appearance, they are known as the *septum primum* and the *septum secundum*. As early as the third to fourth embryonic week, the primordium of the septum primum is laid down as a crescentic ridge on the dorsocephalic part of the atrial wall (Fig. 17). The septum then grows downward, toward the *atrioventricular canal*

At about the same time as the septum primum is laid down, the *atrioventricular canal* also starts to be divided into a right and a left portion (402). A local thickening of the embryonic connective tissue, the *endocardial cushions* (Figs. 17 and 18), is seen on both the dorsal and the ventral wall of the as yet *common canal*. These cushions increase rapidly in size and come in contact with each other about the seventh week. When they become fused, the atrioventricular canal is divided into two entirely separate halves. By the time this fusion is completed, the septum primum has grown to such an extent that it completely separates the two atria, except in a limited area immediately above the cushions, the *interatrial foramen primum* or *ostium primum* (Fig. 19). This foramen decreases successively in size and gradually disappears altogether. Before this occurs, an opening appears in the cephalic part of the septum primum—the *interatrial foramen secundum* (Fig. 19). It arises on

with valves, is developed in a relatively short space of time. It is capable of maintaining the circulation through two separate systems (101, 102, 181, 337, 526, 646).

The *truncus arteriosus* is given off from the cranial part of the aforementioned tube, and the great veins empty into its caudal part (Fig. 13). A factor which greatly contributes to the transformation of this tube

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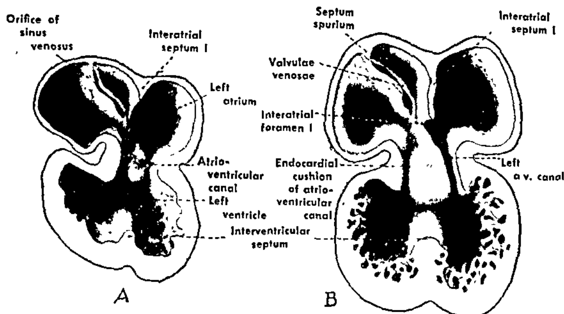


Fig. 17.—Semischematic drawings of inferior aspect of heart to show initial steps in its partitioning. *A*, cardiac septa represented at stage reached in human embryos early in fifth week. Note especially the primary relations of interatrial septum primum Based on original reconstructions of the heart of a 3 7 mm pig embryo, and on Tandler's reconstructions of corresponding stages of the human heart *B*, cardiac septa as they appear in human embryos in the sixth week Note re-

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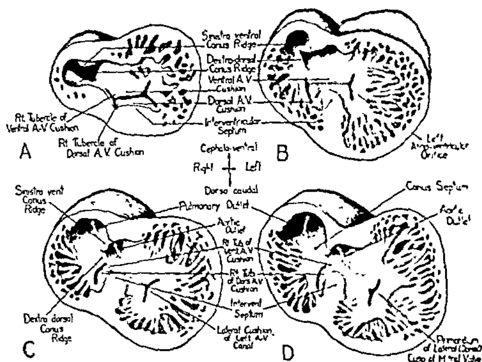


Fig 18 —Four stages in partitioning of atrioventr...
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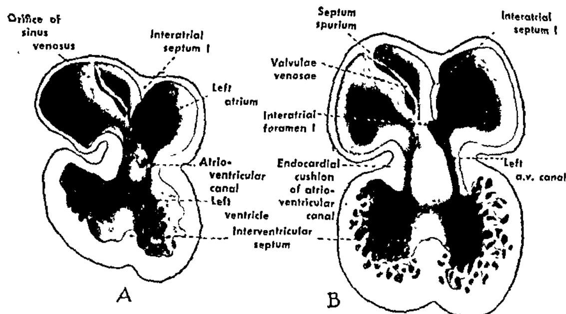
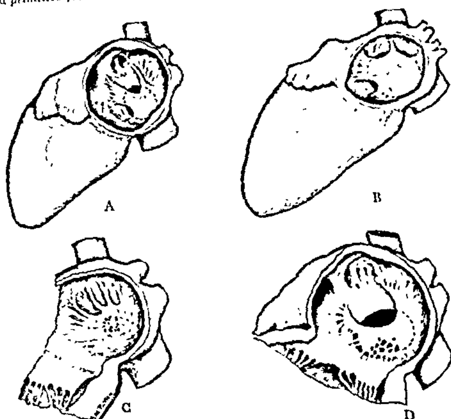


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Even at an early stage, small grooves appear on the external aspect of the heart tube, marking its four different sections the *conus (bulbus cordis)*, the *ventricle*, the *atrium* and the *sinus venosus* (Fig. 13). During the formation of the cardiac loop, these *grooves* become deeper, and at the same time a new groove appears in the medial segment of the *apical part* of the ventricular loop (Fig. 15). This groove, which can sometimes be seen distinctly in the fully developed heart (Fig. 16), indicates the first division of the heart into a left and a right side. There is a concurrent marked increase in the size of both the atrium and the ventricle The atrium enlarges to such an extent that it bulges on either side of the truncus arteriosus and the conus Development then proceeds rapidly, and at the end of the first embryonic month it is so far advanced that the final division of the heart into four chambers is discernible From this stage onward, it is no longer possible to follow the development of the heart as a unit, but each of its parts must thereafter be dealt with separately.

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ig 20.—Various types of defects in valvular mechanism of foramen ovale which prevent tive closure A, defect due to over-resorption of septum primum (stillborn, autopsy 176,204. Inst. ... [illegible]

atrial communication is necessary as as the fetal circulation exists.

he resorption of the septum primum lly starts in the form of small perfora-, which gradually become confluent form a single large opening, cephalo-ally in the septum. Similar small per-ions may occasionally appear at other in the septum and persist without

About simultaneously with the development of the interatrial foramen secundum, a new septum, the *septum secundum*, starts to be formed beside and directly to the right of the septum primum (Fig. 19). Like the septum primum, it is crescentic and sweeps downward and parallel to it like a curtain, toward the opening of the inferior vena cava (434). As the septum

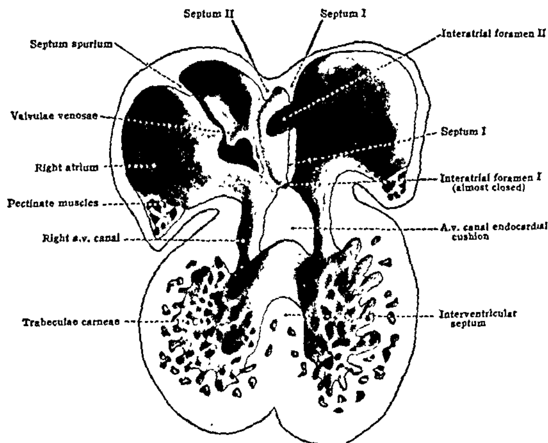


Fig. 19.—Semischematic drawing of interior of heart to show start of interatrial septu

Blakiston Company, 1946])

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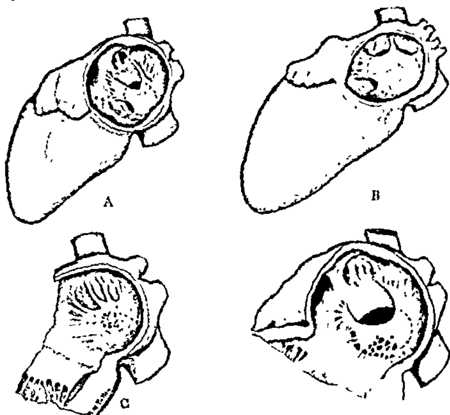


Fig 20 —Various types of defects in valvular ctive closure A, defect due to over-resorption of sep h Inst, Vienna) B, defect due to underdevelopment lly large foramen ovale (female, lived 16 hours, autopsy ated valvula, resorption in abnormal locations (male, t Vienna) D, extensive valvular defect involving a nuoned factors (specimen 4093, Rokitansky Museum, d about 5 months) (From Patten, Am. J. Path 14,135,

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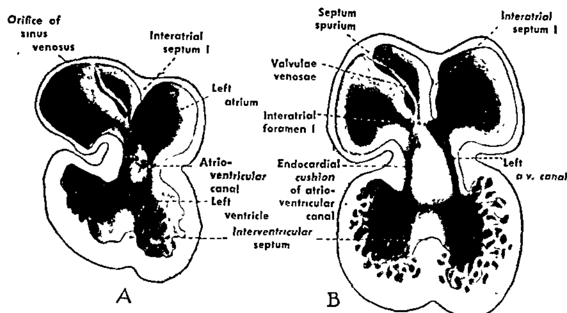


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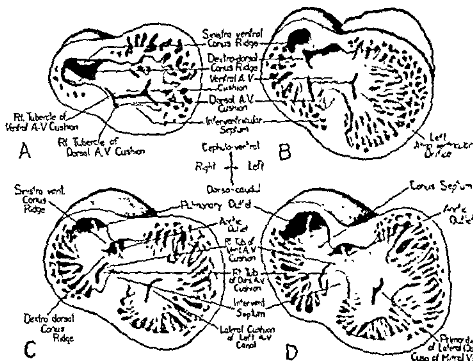


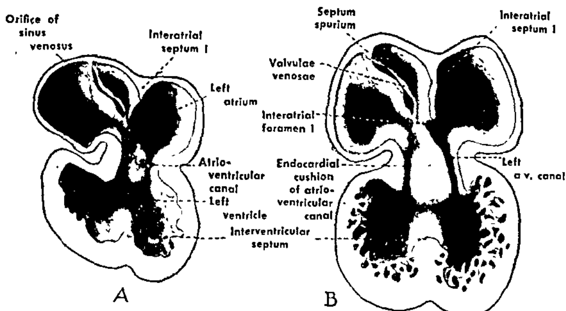
Fig 18.—Four stages in partitioning of atrioventricular canal. The apex of the ventricle has been cut off and the heart is viewed from below. In this aspect, the relations of conus ridges to A-V canal cushions and to upper part of interventricular septum are especially instructive. (From Kramer, Am J Anat. 71 343, 1942.)

of the two atria is effected by the growth of two septa, in view of the time of their appearance, they are known as the *septum primum* and the *septum secundum*. As early as the third to fourth embryonic week, the primordium of the septum primum is laid down as a crescentic ridge on the dorsocephalic part of the atrial wall (Fig 17). The septum then grows downward, toward the *atrioventricular canal*

At about the same time as the septum primum is laid down, the *atrioventricular* entirely separate halves By the time this fusion is completed, the septum primum has grown to such an extent that it completely separates the two atria, except in a limited area immediately above the cushions, the *interatrial foramen primum* or *ostium primum* (Fig. 19). This foramen decreases successively in size and gradually disappears altogether. Before this occurs, an opening appears in the cephalic part of the septum primum—the *interatrial foramen secundum* (Fig. 19). It arises on

with valves, is developed in a relatively short space of time. It is capable of maintaining the circulation through two separate systems (101, 102, 181, 337, 526, 646).

The *truncus arteriosus* is given off from the cranial part of the aforementioned tube, and the great veins empty into its caudal part (Fig. 13). A factor which greatly contributes to the transformation of this tube



of the heart of a 3 7 mm pig embryo, and on the human heart. *B*, cardiac septa as they ap

is its rapid increase in length. Owing to lack of space, it is at first bent sideways, but subsequently curves in a pronounced loop (Figs 14 and 15) Since the cephalic part of the heart is stabilized by the truncus arteriosus and the caudal part by the great veins, the middle part undergoes the greatest changes in position.

Even at an early stage, small grooves appear on the external aspect of the heart tube, marking its four different sections the *conus (bulbus cordis)*, the *ventricle*, the *atrium* and the *sinus venosus* (Fig. 13). During the formation of the cardiac loop, these *grooves* become deeper, and at the same time a new groove appears in the medial segment of the apical part of the ventricular loop (Fig. 15). This groove, which can sometimes be seen distinctly in the fully developed heart (Fig. 16), indicates the first division of the heart into a left and a right side. There is a concurrent marked increase in the size of both the atrium and the ventricle. The atrium enlarges to such an extent that it bulges on either side of the truncus arteriosus and the conus. Development then proceeds rapidly, and at the end of the first embryonic month it is so far advanced that the final division of the heart into four chambers is discernible From this stage onward, it is no longer possible to follow the development of the heart as a unit, but each of its parts must thereafter be dealt with separately.

1. The Atria and the Atrioventricular Canal

The actual division of the common atrium into two chambers takes place during the second fetal month. Complete separation does not, however, occur until after birth, when the placental circuit has been cut off and the pulmonary circulation has become stabilized. The internal separation canal also starts to be divided into a right and a left portion (402). A local thickening of the embryonic connective tissue, the *endocardial cushions* (Figs. 17 and 18), is seen on both the dorsal and the ventral wall of the as yet *common canal.* These cushions increase rapidly in size and come in contact with each other about the seventh week. When they become fused, the atrioventricular canal is divided into two

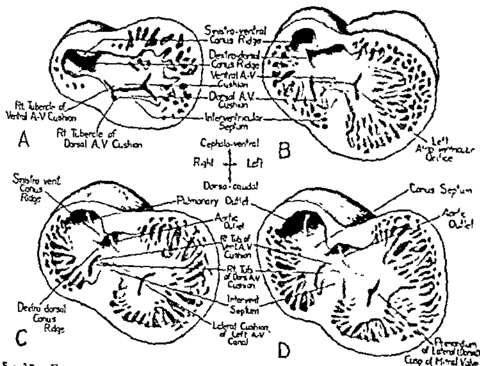


Fig. 18.—Four stages in partitioning of atrioventricular canal. ... been cut off and the heart is viewed fro... A-V canal cushions and to upper part of ... Kramer, Am. J. Anat. 71: 343, 1942.)

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the...

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entirely separate halves. By the time this *fusion* is completed, the septum primum *has grown* to such an extent that it completely separates the two atria, except in a *limited area* immediately above the cushions, the *interatrial foramen primum* or *ostium primum* (Fig. 19). This *foramen* decreases successively in size and gradually disappears altogether. Before *this* occurs, an opening appears in the *cephalic part* of the septum primum—the *interatrial foramen secundum* (Fig. 19). It arises on

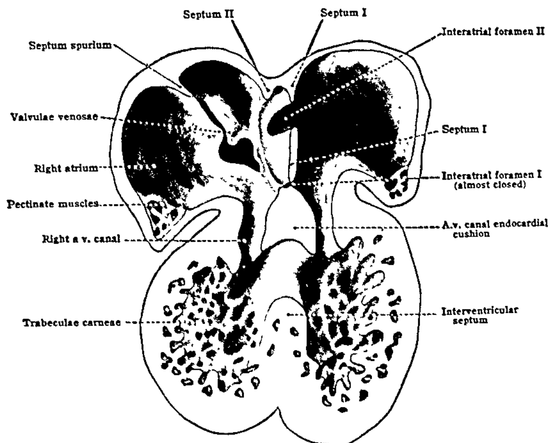


Fig. 19.—Semischematic drawing of interior of heart to show start of interatrial septu

Blakiston Company, 1946].)

the grounds of resorption of a limited area of the septum. This opening enlarges and, owing to the concomitant growth of the septum primum, is displaced somewhat ventrally and caudally (102). It forms the so-called *primitive foramen ovale*. Such an being incorporated with the foramen secundum (521) (Figs. 20 and 378, p. 410). If these openings lie directly opposite the foramen ovale in the septum secundum, they create the possibility of an interatrial shunt postnatally.

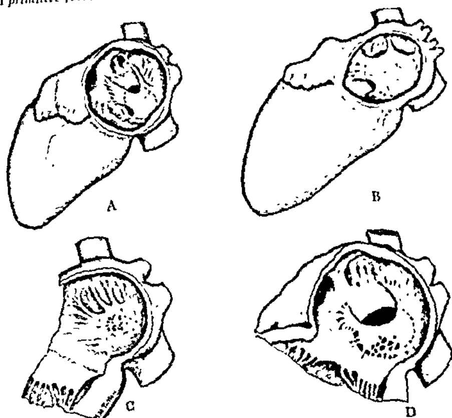


Fig. 20.—Various types of defects in valvular mechanism of foramen ovale which prevent

interatrial communication is necessary as long as the fetal circulation exists.

The resorption of the septum primum usually starts in the form of small perforations, which gradually become confluent and form a single large opening, cephalodorsally in the septum. Similar small perforations may occasionally appear at other sites in the septum and persist without

About simultaneously with the development of the interatrial foramen secundum a new septum, the *septum secundum* starts to be formed beside and directly t the right of the septum primum (Fig. 19) Like the septum primum, it is crescenti and sweeps downward and parallel to like a curtain, toward the opening of th inferior vena cava (434). As the septu

secundum, which is considerably thicker than the septum primum, approaches the entry of the inferior vena cava, its growth decreases and ceases entirely before reaching this opening. Here it leaves a characteristic oval gap, the *foramen ovale*, which has a thickened superior border, the *limbus fossae ovalis* (Fig. 21). The direction of the foramen ovale is such that the greater part of the blood flow from the inferior vena cava is routed directly into the left atrium. The position of the foramen ovale does not correspond to the position of the aforementioned foramen secundum, it lies caudally and dorsally to it It is therefore entirely covered by the thin septum primum (528) (Fig. 22)

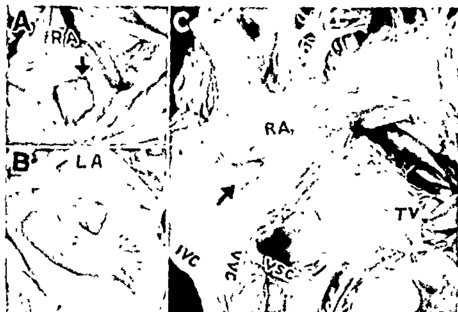
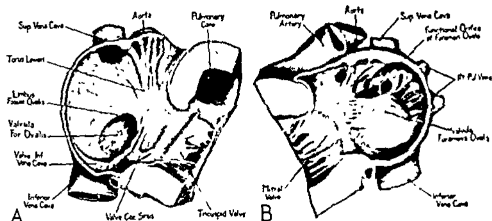


Fig. 21.—Closed foramen ovale *A*, from right atrium (the limbus fossae ovalis is prominent); *B*, from left atrium, *C*, very conspicuous limbus. *IVC*, inferior vena cava, *LA* and *RA*, left and right atria; *TV*, tricuspid valve; *VSC*, valvula sinus coronarii, *VVC*, valvula venae cavae, *arrow in A points to limbus fossae ovalis, arrow in C points to foramen ovale*

Since no fusion takes place between the septa in the limbus region, the septum secundum acts as a one-way valve (Fig 22) When the fetal circulation ceases, the function of the valve ends and the process of closure of the foramen ovale can start The connective tissue of the valve then begins to proliferate, and when this process is optimal, an adhesion of the valve takes place, so that it becomes an integral part of the atrial septum (Figs. 23 and 24). There is a wide range of variation in the time for final closure of the foramen ovale (577). Closure before 3 months of age is unusual, and, according to Patten (523), it commonly occurs in the last third of the first postnatal year. In 20 to 25 per cent of all individuals (312, 391 (in 33 per cent), the fibrous fusion is never complete (Fig. 25).

Like the septum primum, the septum secundum grows downward and fuses with the cushions of the atrioventricular canal. Patten (527) has stated that if the cushions fail to fuse, both the septum primum and septum secundum tend to be underdeveloped in this region. An opening at the site at which they should fuse with the cushions then persists (Fig 26) and is commonly called a *persistent interatrial foramen primum.*

According to Pfenning (536) and Faber (246), among others, the septum primum or secundum may be laid down at an abnormal site in the atrium. The two septa then run entirely or partly separate from each other, and a *third atrium* is delimited between them. *A third atrium may also*



the functional orifice is considerably smaller than the oval opening in the septum secundum (From Patten, Sommerfield and Paff, Anat. Rec. 44:165, 1929)

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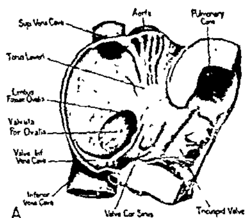
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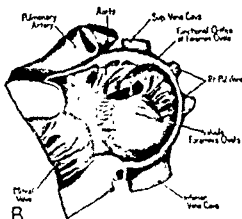
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Interior views of scale model of the human heart at term. A, right atrial aspect show

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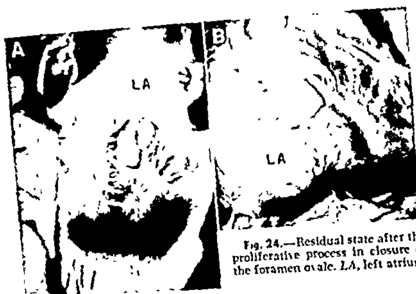


Fig. 24.—Residual state after the proliferative process in closure of the foramen ovale. *LA*, left atrium.

A
LA
MV
B
TV
IVC
C
LA
D
SVC
VVC
TV
IVC

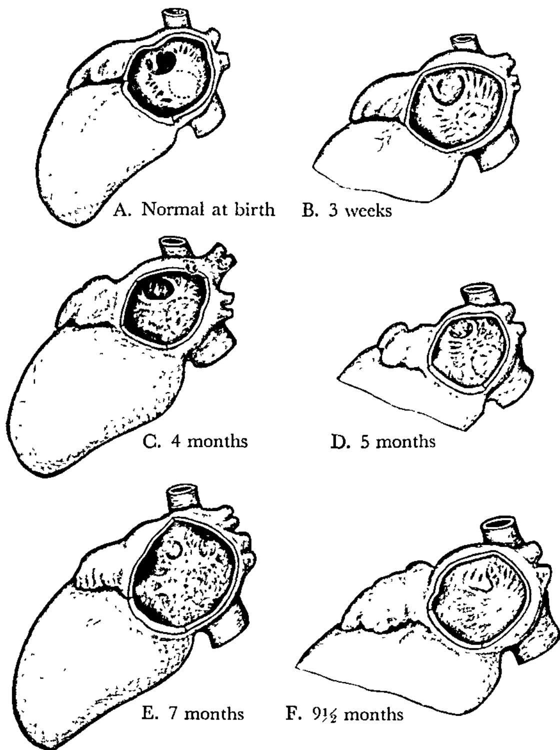


Fig. 23.—Drawings of hearts with left atrium opened to show gross changes in valvula during period of closure of foramen ovale (From Patten, Am J Anat 48 19, 1931)

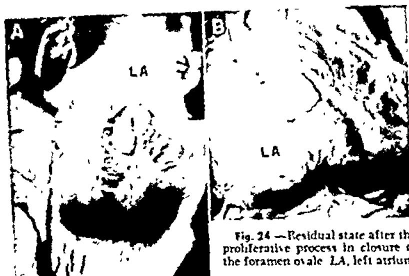
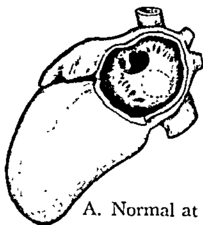
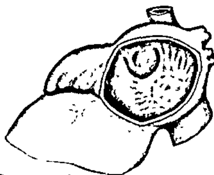
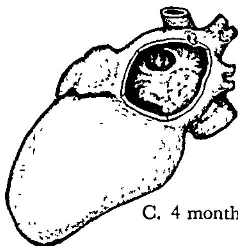


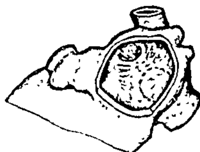
Fig. 24.—Residual state after the proliferative process in closure of the foramen ovale. *LA*, left atrium.

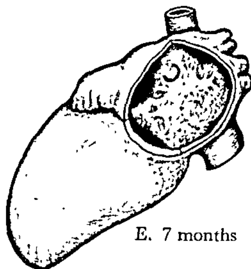
Fig. 25.—Patent foramen ovale. *A* and *C*, left atrium, probe in the foramen ... right atrium. In *B*, the ...











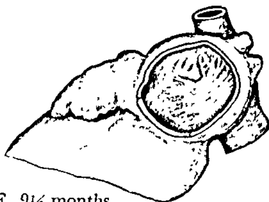
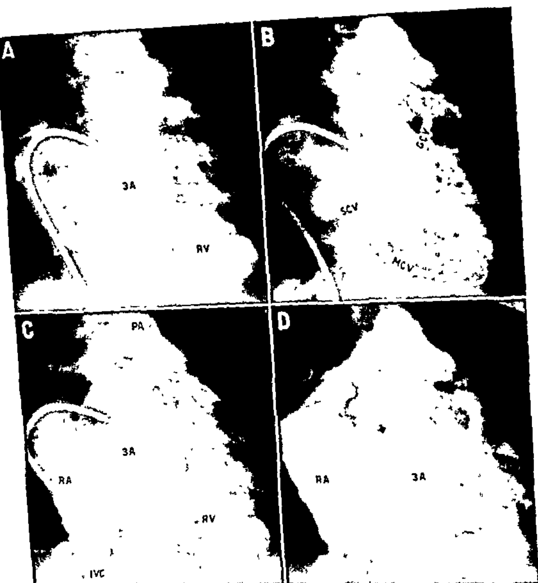


Fig. 23.—Drawings of hearts with left atrium opened to show gross changes in valvula during period of closure of foramen ovale (From Patten, Am J Anat. 48 19, 1931)



[illegible]

IVC, inferior vena cava, *MCV*, middle cardiac vein, *PA*, pulmonary artery, *RA*, right atrium, *RV*, right ventricle (*continued*)

arise as a result of other anomalous septal formations. Opinions differ greatly with regard to their pathogenesis. Depending on the position of the abnormal septum, different groups of veins open into the third atrium (211, 257, 522, 531, 606). The most common variant is a division of the left atrium into two parts, *double left atrium* (222, 430, 535, 630, 672), with a varying number of pulmonary veins emptying into the respective halves. A similar division of the right atrium is occasionally found (Fig 27). Here, a third atrium—into which an abnormally situated coronary sinus opens —is seen between the right and the left atrium. It may be inferred from Figure 27 that the course of the cardiac veins also differs appreciably from the normal course.

A *pouchlike dilatation of the proximal parts of one or several pulmonary veins* (Fig. 28), reminiscent of a third atrium, is sometimes seen. This dilatation usually arises on the basis of constriction of the orifice (224).

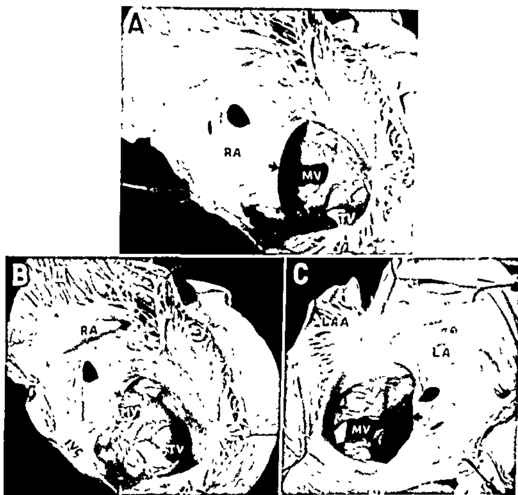


Fig. 26.—Persistent interatrial foramen primum, underdevelopment of septum secundum. with patent foramen ovale and resorption at an abnormal site in the septum primum *IVC*, inferior vena cava; *LA* and *RA*, left and right atria, *MV*, mitral valve, *TV*, tricuspid valve, arrow points to foramen primum, *LAA*, left auricular appendage.

Concurrently with the beginning development of the septum primum and the endocardial cushions of the atrioventricular canal, the *sinus venosus* is shifted from the midline, so that it opens entirely into the future right atrium, to the right of the newly formed atrial septum (Fig 19) The orifice of the sinus venosus is guarded by two well developed valves, the *right and left venous valves* (Figs. 29 and 30). At

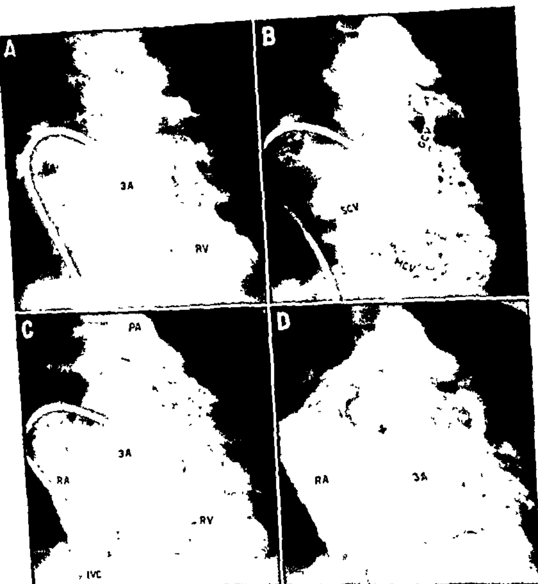


Fig 27.—Infundibular pulmonary stenosis Boy, aged 6 (K D 470602). During injection, the tip of the catheter recoils from right ventricle into the right atrium The jet of contrast medium is then directed to the left, and filling is obtained of a fairly sharply delimited cavity lying between [illegible]

right ventricle (*continued*)





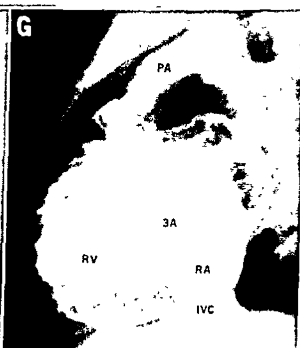


Fig 27 (*cont.*)



Fig. 28 —Abnormal venous return Boy, aged 8 (A R. 470325). Septum seen (arrow in *E*) ulging into the lumen at the level of the opening of the right superior pulmonary vein (arrows n F). *AO* and *Ad*, ascending and descending aortae, *LA*, left atrium, *LV*, left ventricle, *PV*, pulmonary vein.

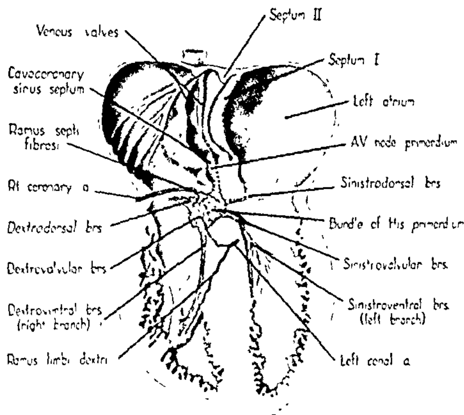


Fig. 29.—Semischematic diagram to show the branches of the coronary arteries supplying the bundle of His and its main branches The positional relation of the venous valves to the atrial septum is clearly evident The illustration was drawn as if the composite mass of endocardial tissue which occludes the interventricular foramen had been completely removed to expose the bundle of His (From Licata, Am J Anat. 94:73, 1954.)

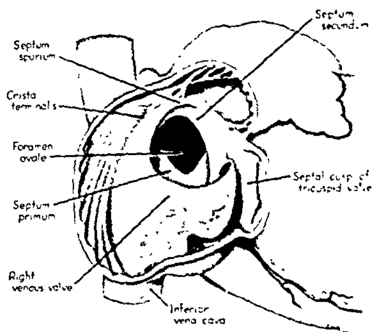


Fig 30a.—Dextral view of heart of a 31.5 mm embryo, with the right atrium opened The lower part of the right venous valve shows local areas of resorption. (From Licata, Am J. Anat. 94 73, 1954)

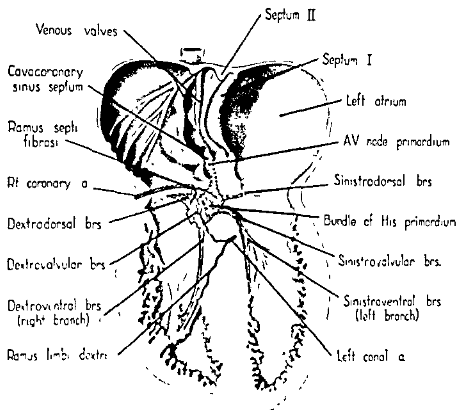


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he cephalic end they are continuous with a powerful, ridgelike fold, the *septum spurium* (Fig. 19), which takes its origin from the cephalic wall of the atrium (434, 510). According to Schmidt (580), the right valve attains a considerable size in the third fetal month. Because of its position, in front of the orifices of the superior and inferior venae cavae, it helps to direct the blood from these vessels into the proper chamber of the heart. The septum spurium, as well as the right and left venous valves, gradually undergo considerable resorption, but the two first-mentioned are not entirely resorbed In the adult, the persistent part of the septum spurium is represented by the *crista terminalis* (Figs 65 and 66, pp. 65 and 66) The remains of the right venous valve from the *eustachian valve* of the inferior vena cava and the *thebesian valve* of the coronary sinus (Figs 73 and 74, p 73) Occasionally (312) in 3.2 per cent of all cases, the septum spurium and venous valves are resorbed to a lesser degree and leave a network of bands, Chiari's net (Fig. 31), attached to the crista terminalis and the margins of the eustachian and thebesian valves.

A defect which, cephalad, lacks a complete margin (see p. 34) is sometimes found high up in the atrial septum, just below the superior vena cava. The embryologic development of this defect is not yet fully elucidated. According to Ross (568) there is good evidence from a consideration of the embryology that it results from a developmental defect of the sinus venosus. Hudson (350) explains the defect as a persistence of the vestibule of the sinus venosus In the ordinary course of events, this sinus is absorbed into the posterior wall of the right atrium. According to Keith (378), in the fetal stage an extension is found laterally behind the left atrium, as the vestibule of the sinus venosus, into which the pulmonary veins empty.

In man, this extension toward the left side of the common atrium normally be-



Fig 31 (*left*) —Chiari's net, which extends from the region above the superior vena cava (*SVC*) to the inferior vena cava (*IVC*)

Fig 32 (*right*) —Beside the margin of the closed foramen ovale are narrow strands and bands, presumably remnants of the left valve of the sinus venosus *FoO*, fossa ovalis, *RA*, right atrium.

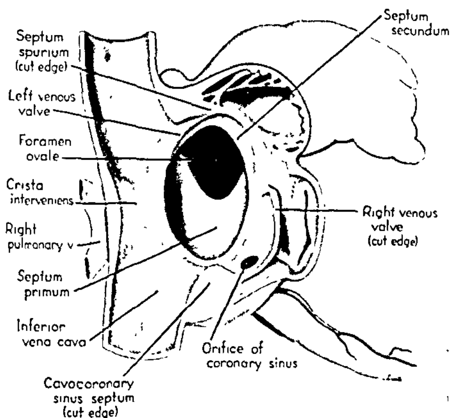


Fig. 30b.—Dextral view of heart of 31 5 mm embryo, with the right atrium opened and right venous valve removed. (From Licata, Am. J. Anat. 94.73, 1954.)

the cephalic end they are continuous with a powerful, ridgelike fold, the *septum spurium* (Fig. 19), which takes its origin from the cephalic wall of the atrium (434, 510). According to Schmidt (580), the right valve attains a considerable size in the third fetal month. Because of its position, in front of the orifices of the superior and inferior venae cavae, it helps to direct the blood from these vessels into the proper chamber of the heart. The septum spurium, and leave a network of bands, Chiari's net (Fig 31), attached to the crista terminalis and the margins of the eustachian and thebesian valves.

A defect which, cephalad, lacks a complete margin (see p 34) is sometimes found high up in the atrial septum, just below the superior vena cava. The embryologic development of this defect is not yet fully elucidated According to Ross (568) there is good evidence from a consideration



Fig. 31 (*left*) —Chiari's net, which extends from the region above the superior vena cava (*SVC*) to the inferior vena cava (*IVC*)

Fig. 32 (*right*) —Beside the margin of the closed foramen ovale are narrow strands and bands, presumably remnants of the left valve of the sinus venosus *FoO*, fossa ovalis, *RA*, right atrium.

as well as the right and left venous valves, gradually undergo considerable resorption, but the two first-mentioned are not entirely resorbed In the adult, the persistent part of the septum spurium is represented by the *crista terminalis* (Figs 65 and 66, pp. 65 and 66). The remains of the right venous valve from the *eustachian valve* of the inferior vena cava and the *thebesian valve* of the coronary sinus (Figs 73 and 74, p 73) Occasionally (312) in 3 2 per cent of all cases, the septum spurium and venous valves are resorbed to a lesser degree of the embryology that it results from a developmental defect of the sinus venosus. Hudson (350) explains the defect as a persistence of the vestibule of the sinus venosus In the ordinary course of events, this sinus is absorbed into the posterior wall of the right atrium. According to Keith (378), in the fetal stage an extension is found laterally behind the *left atrium*, as the vestibule of the sinus venosus, into which the pulmonary veins empty.

In man, this extension toward the left side of the common atrium normally be-

comes cut off from the main sinus venosus by the interatrial septum, so that the pulmonary veins come to open into the left atrium. In those cases in which absorption of the sinus venosus into the atrium is incomplete, a persistent communication with the vestibule arises, adjacent to the mouths of the superior vena cava and pulmonary veins. Licata (434) states that the regression of the *left venous valve* becomes increasingly closely related to the septum secundum. In the beginning of the ninth week, a cleft-like recess is present between the superior part of the left venous valve and the interatrial septum secundum (Figs. 29 and 30). This recess is a remnant of an *originally wider atrial pocket* In earlier stages, the cephalic limit of the left venous valve could be clearly identified as the point at which it met the right venous valve to form the septum spurium. By nine weeks, however, regression of the most cephalic portion of the septum spurium is well advanced. What remains of it has, for the most part, become merged with the septum secundum, so that considerable obliteration of the space has occurred. The upper part of the left venous valve as well is involved in the fusion. As a consequence, it has become *directly adherent* to the developing septum secundum in this region. Thus it is conceivable that the left venous valve partakes as an *integrating part in* formation of the atrial septum and division of the atrium. Consequently, excessive regression of this valve should then result in a persisting communication between the atria.

The *left valve of the sinus venosus* usually disappears entirely, but small remnants of its superior part may exceptionally persist in the form of small strands adherent to the septum secundum, usually close to the foramen ovale (Fig. 32). In rare cases, *strands* may also be found in the *left atrium;* they generally issue from the foramen ovale region and represent rudimentary equivalents of the septum formations which contribute to division of the atrium (Fig. 33)

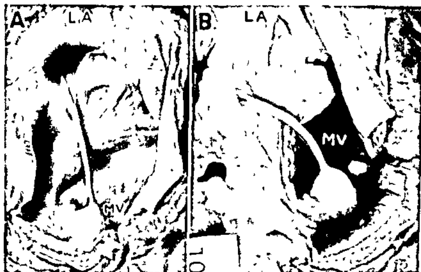


Fig. 33.—Rudimentary septum formations in the left atrium (*LA*) *MV*, mitral valve

2. The Ventricular Septum

At about the same time as the first atrial septum is laid down, the initial signs appear of division of the ventricle into a right and a left half. During its initial stage, the *ventricular septum* is indicated by a small ridge *with a reticular appearance. According* to Streeter (634), it is formed from the *trabeculae carneae* present along the ventricular wall (Fig. 34) As development progresses, the trabeculae fuse into a relatively solid mass. This fusion may

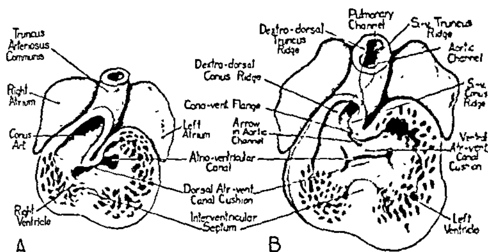


Fig. 34.—Semischematic dissections of developing heart viewed from frontal aspect to show relations of importance in establishing aortic and pulmonary outlets. (From Kramer, Am. J. Anat 71 343, 1942.)

sometimes be incomplete, and small *interventricular defects* persist. They are invariably situated in the muscular part of the septum (Fig. 35).

According to current conceptions, true growth of the septum takes place in the direction of the cushions of the atrioventricular canal. Patten (527) has nevertheless stated that the actual course of events is that, with the growth of the ventricles on either side of the septum, the septum bulges to a relatively increasing extent into the wide ventricles Consequently, the *interventricular foramen* appears to become relatively smaller but actually is almost unchanged

The closure of the interventricular foramen is an exceedingly complex procedure and affords the possibility of several malformations. It takes place by means of closely interdependent processes in various endocardial cushion tissue masses. They are derived from the conus ridges and the crest of the ventricular septum, as well as from the tubercles which develop from the endocardial cushions of the atrioventricular canal (261, 402, 511) (Fig 36) The muscular part of the septum does not, however, take part in this process of closure, which is usually completed by the end of the second embryonic month. Here development of the conus ridge is significant.

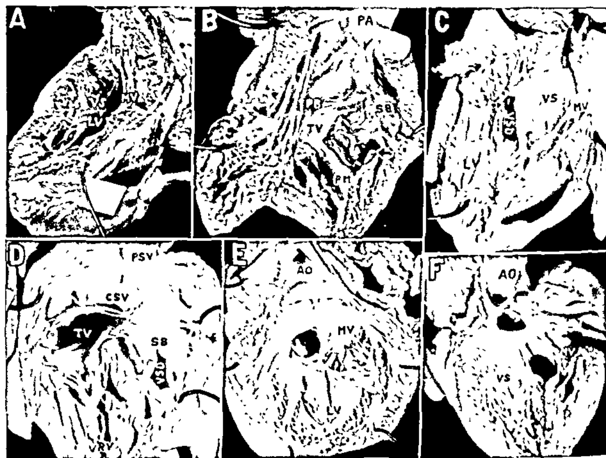


Fig. 35.—Defects in muscular part of the ventricular septum *A*, left ventricle, *B*, right ventricle, with probe in the defect, *C*, left ventricle, *D*, right ventricle, *E*, defect high in the muscular part of the septum, *F*, defects in both membranous and muscular parts of the septum *AO*, aorta, *CSV*, crista supraventricularis, *I*, infundibulum, *LV* and *RV*, left and right ventricles, *MV*, mitral valve, *PA*, pulmonary artery, *PB*, parietal band, *PM*, papillary muscle, *PSV*, pulmonary semilunar valve, *SB*, septal band, *TV*, tricuspid valve, *VS*, ventricular septum, *VSD*, ventricular septal defect.

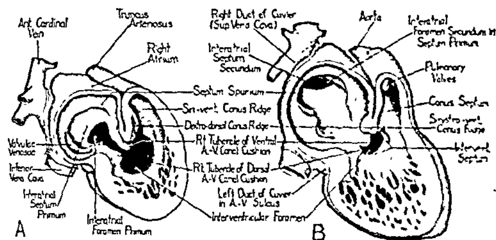
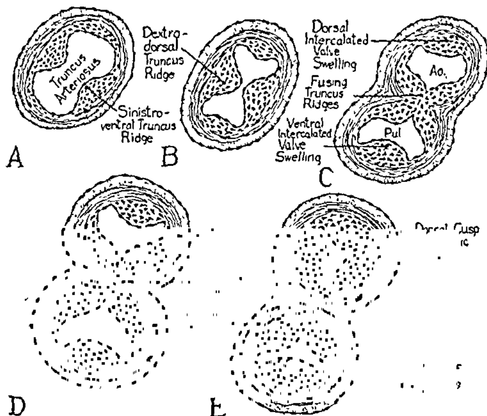


Fig 36.—Schematic lateral dissections to show relations of various septa in the developing eart (From Patten, B M.: Human Embryology [New York: Blakiston Company, 1946])

3. Development of the Conus Septum and Truncus Arteriosus

In the early stage of fetal development, *the conus denotes the common outflow region of both future ventricles.* It is separated from the left ventricle by a deep groove, the *conoventricular sulcus* (Fig. 34, *A*), to which a large *flange* corresponds on the inner surface of the heart. This flange contributes to the passage of blood from the left ventricle wholly or partly through the right ventricle before it passes into the conus and the truncus arteriosus. Concurrently with the beginning division of the conus and truncus arteriosus and the continued expansion of the ventricles, this flange begins to diminish in size and gradually disappears almost entirely (Fig. 34, *B*).

The first sign of separation of the *conus and the truncus arteriosus* is the appearance of two *longitudinal ridges* of connective tissue, placed on either side of the inner wall (Fig. 37). They pursue a clockwise spiral course—a fact that is probably *dependent on the twisting of the* aorta and the pulmonary artery around each other—from the origin of the sixth aortic arches, through the truncus and conus toward the ventricular septum. According to Kramer (402), these ridges do not develop uniformly, but some parts grow more rapidly than others This applies in particular at the site of origin of the sixth aortic arches and at the future site of the semilunar valves. These ridges gradually meet in the midline and fuse. They then delimit an *aortic channel*, which joins the left ventricle with the fourth pair of aortic arches, and a *pulmonary channel*, which forms the communication between the right ventricle and the sixth pair of aortic arches If the formation of these ridges in the truncus



arteriosus does not take place, no division of this vessel occurs, and a true persistent truncus arteriosus arises (Fig. 38).

If development is incomplete, a *communication between the aorta and the pulmonary artery* persists, in the form of a round



Fig. 38.—Persistent truncus arteriosus. The pulmonary artery arises from the posterior aspect of the truncus, about 4 cm above the valvular plane. The opening to the ...
the pu
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or oval hole, slightly above the valves, in the anterior wall of the aorta. If one vessel is smaller than the other, this can be explained hypothetically on the grounds of unequal division of the truncus. The difference between the caliber of the vessels can, however, be entirely a result of the flow, as has been pointed out by Shaner (598), among others (Fig. 39). If either of these ridges is not laid down in the conus region, or if they develop incompletely or at an anomalous site, a *defect between the arterial and the venous part of the conus* develops, as shown in Figure 48. Small remains persisting after fusion of the ridges can sometimes be observed at autopsy. This applies particularly when fusion has been incomplete (Fig. 40).

Slightly before fusion of the ridges, small cushions of connective tissue are visible on the inner surface of both the aorta and the pulmonary artery, at the junction of the truncus arteriosus and the conus (Fig. 37, C–E). During their growth, these cushions are modified into the *cusps of the semilunar valves*. Maldevelopment can result in the cusps being absent (141) or two in number (Fig. 41), four (Fig. 42) (483), or, in rare cases, more. In this event, there are not infrequently associated malformations.

In this connection, Shaner's interesting embryologic investigations (598, 599) should be mentioned. Although they are perhaps not directly applicable in man, they throw some light on the way in which an early malformation of the endocardial cushions of the atrioventricular canal can



Fig. 39.—Tricuspid ...

give rise to deformations of the truncus region.

In examination of 15,000 pig embryos, Shaner observed a definitely abnormal heart in 35 cases. In 16 of them, there was

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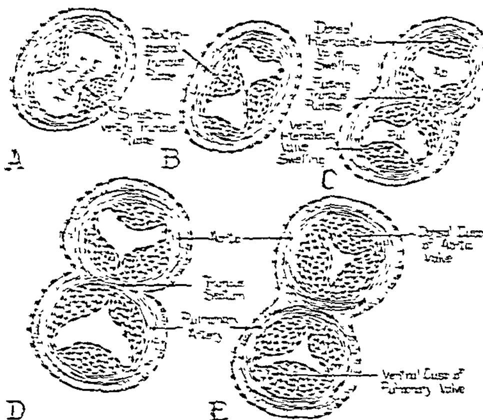


Fig. 37.—Schematic cross-sectional diagrams to explain partitioning of truncus arteriosus. Relations are depicted as they would appear at the level of the developing semilunar valves. (From Kramer: Am. J. Anat. 71: 343, 1942.)

The first sign of separation of the *conus* and the *truncus arteriosus* is the appearance of two *longitudinal ridges* of connective tissue, placed on either side of the inner wall (Fig. 37). They pursue a clockwise spiral course—a fact that is probably dependent on the twisting of the aorta and the pulmonary artery around each other—from the origin of the sixth aortic arches through the truncus and conus toward the ventricular septum. According to Kramer (432), these ridges do not develop uniformly, but some parts grow more rapidly than others. This applies in particular at the site of origin of the sixth aortic arches and at the future site of the semilunar valves. These ridges gradually meet in the midline and fuse. They then delimit an *aortic channel*, which joins the left ventricle with the fourth pair of aortic arches, and a *pulmonary channel*, which forms the communication between the right ventricle and the sixth pair of aortic arches. If the formation of these ridges in the truncus

arteriosus does not take place, no division of this vessel occurs, and a true persistent truncus arteriosus arises (Fig 38).

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Fig. 38 —Persistent truncus arteriosus. The pulmonary artery arises from the posterior aspect of the truncus, about 4 cm above the valvular plane. The opening to the right branch of the pulmonary artery is visible immediately beside its origin from the truncus. *AO*, aorta, *PA*, left branch of pulmonary artery, *Tr*, truncus

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Fig 39.—*Tricuspid* atresia, patent foramen ovale, rudimentary right ventricle with closed ventricular septum, pulmonary *atresia* and patent ductus arteriosus. Girl, aged 14 days (Y.W. 540101). Lungs are … mainly thr… (*PDA*) of a… nary artery … *A* and *RPA*, left and right pulmonary arteries.

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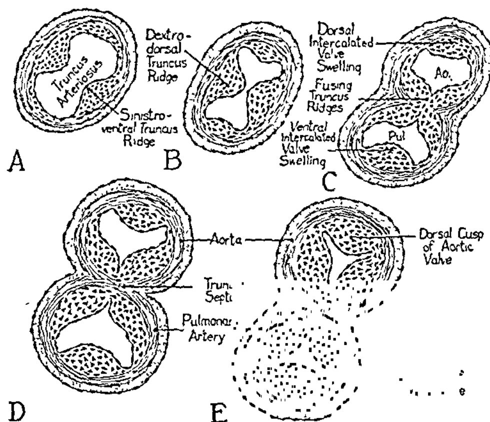


Fig. 37.—Schematic cross-sectional diagrams to explain partitioning of truncus arteriosus Relations are depicted as they would appear at the level of the developing semilunar valves (From Kramer, Am. J. Anat. 71.343, 1942)

malformation of the conus and great vessels, combined with defective fusion of the atrioventricular canal cushions Moreover, the aorta was over-riding or opened entirely into the right ventricle, and the aorticopulmonary septum was defective or the whole of its proximal part was lacking.

According to Shaner, the conus and truncus arteriosus in a 6 mm pig embryo consist of a simple tube, taking its origin entirely from the right ventricle (Fig 43, 1) Between the two ventricles is a powerful conoventricular ridge. In a later stage, the truncus arteriosus becomes divided into the aorta and pulmonary artery. The vessel orifices migrate to the left, so that the transposed. Should the aorta nevertheless continue to migrate, it is pushed forward between the abnormal, ventral atrioventricular cushion and the ventricular wall. Retracted migration of the aortic orifice results in an abnormal position of the aorticopulmonary septum, which, in turn, prevents closure of the ventricular septum. Doerr (198) now supports this conception in man

In three of Shaner's nine cases described, the aorta and the pulmonary artery were of the same caliber, whereas aortic stenosis was present in three cases and pulmonary stenosis in three. Shaner has stated that if the blood supply to the aorta and pul-



Fig. 42.—Pulmonary valve with four cusps *PA*, pulmonary artery

pulmonary orifice lies directly beside the ventricular septum, and the aortic orifice, just medial to the aortic cusp of the mitral valve (Fig 43, 5). To permit this migration it is, however, necessary for the following events to take place (1) The conoventricular ridge must disappear almost completely (Fig 43, 2); (2) the ventral cushion must, when the dorsal and ventral atrioventricular canal cushions have become fused (Fig 43, 2), become so greatly excavated in the center that practically only its two tubercles remain (Fig. 43, 3). This is because the aortic channel must pass over the right tubercles of the atrioventricular cushions to reach the left ventricle If the ventral cushions fail to fuse in the normal way with the dorsal cushions and do not become sufficiently deeply excavated, the migration of the aorta is completely arrested or it assumes an abnormal position In the former case, the position of the aortic orifice becomes over-riding or monary artery is nevertheless adequate, their caliber will be normal. If, on the contrary, the blood volume is decreased, the vessel will be narrower, and vice versa.

4. Closure of the Interventricular Foramen

As stated previously, closure of the interventricular foramen takes place by the growth of *endocardial cushions*. These endocardial tissue masses emanate from the conus ridges and the crest of the ventricular septum, and also from the tubercles developed from the endocardial cushions of the atrioventricular canal (Figs. 36 and 44). Although the contribution of the tubercles of the atrioventricular canal cushions is by no means negligible, the two conus ridges are mainly responsible for the closure.

During their growth, the ridges are not confined to the conus region, dividing it



Fig. 40.—Small defects (arrows) in fusion of the conus ridges. When specimen *B* was stretched, a small rupture occurred (slightly below arrow). *PA*, pulmonary artery, *PB*, parietal band, *PM*, papillary muscle; *SB*, septal band, *TV*, tricuspid valve.

Fig. 41.—Bicuspid valves. *A*, aorta, and *B*, pulmonary valve from same patient *AO*, aorta; *MV*, mitral valve, *PA*, pulmonary artery *C* and *D*, bicuspid aortic valve, left cusp is incompletely separated Both coronary arteries arise from the left cusp, one from each corner, that farthest to the left originates in the actual margin of the section *ASV*, aortic semilunar valve, *CA*, coronary artery

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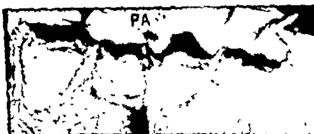


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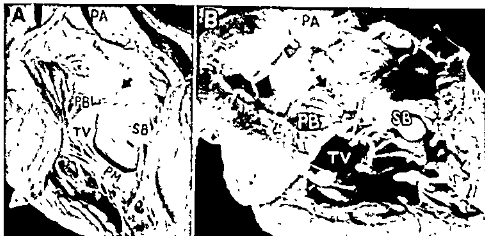


Fig. 40.—Small defects (arrows) in fusion of the conus ridges. When specimen *B* was stretched, a small rupture occurred (slightly below arrow). *PA*, pulmonary artery, *PB*, parietal band; *PM*, papillary muscle, *SB*, septal band, *TV*, tricuspid valve.

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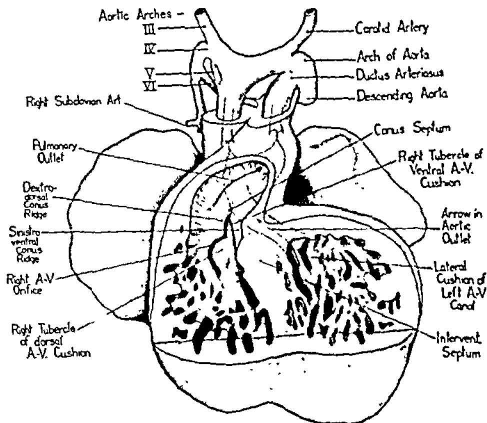


Fig. 44.—Reconstruction of heart of a 13 mm embryo, opened to show relations of conus septa to interventricular septum and the atrioventricular canal cushions. (From Kramer, Am J. Anat. 71 343, 1942.)

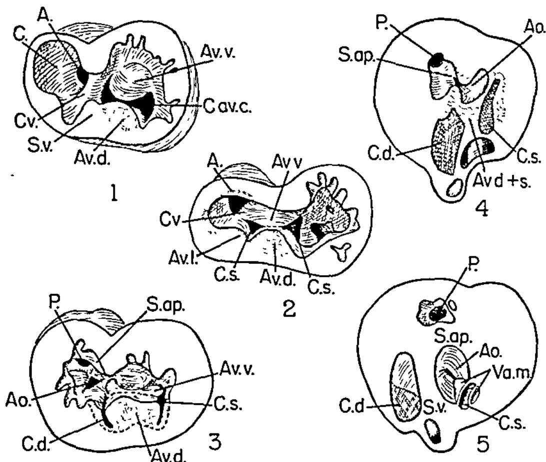


Fig. 43.—Interior views of reconstructions of hearts from normal pig embryos to show normal development of the dorsal and ventral atrioventricular cushions, the resorption of the conoventricular ridge, and the migration of the aortic inlet. The apical part of each model has been removed. The reader looks into the base of the ventricles and into the conus *1*, from a 6 mm embryo, 2, from a 7 mm embryo, 3, from a 10 mm embryo, 4, from a 14 5 mm embryo, 5, from a 27 mm embryo. *A* , truncus arteriosus, *Ao.*, aorta; *Av d ,v.,l.*, dorsal, ventral, and lateral atrioventricular endocardial cushions, *C* , conus, *C av.c* , common atrioventricular canal, *C.d ,s.*, right and left atrioventricular canal, *Cv* , conoventricular ridge, *P.*, pulmonary artery; *S.ap* , aorticopulmonary septum, *S.v* , interventricular septum, *Va m.*, mitral valve. (From Shaner, Am. J. Anat. 84.431, 1949)

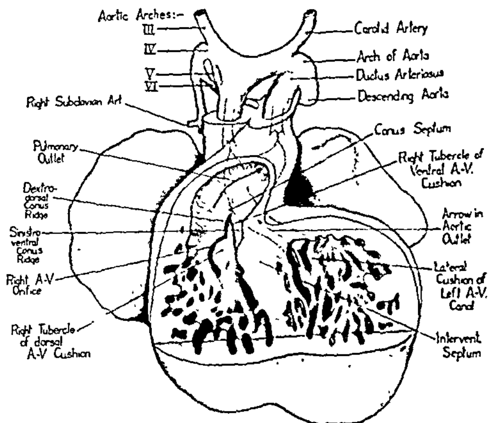


Fig 44.—Reconstruction of heart of a 13 mm embryo, opened to show relations of conus septa to interventricular septum and the atrioventricular canal cushions. (From Kramer, Am J Anat. 71 343, 1942.)

into two halves, but they extend toward the common ventricle, at first in the direction of the ventricular septum. The subsequent growth of the two ridges nevertheless takes place in widely different directions. The *sinistroventral conus ridge* grows downward, forming the *septal band of the crista* *supraventricularis*, along the right side of the ventricular septum, toward the left caudal segment of the anterior ventricular wall, where it merges with its trabeculae (Figs. 36 and 45) The *dextrodorsal conus ridge* curves toward the *orifice of the right atrioventricular canal*. Part of it, the *parietal band of the crista supraventricularis*, continues toward the right caudal part of the anterior ventricular wall, where it joins the trabeculae of this wall. Another part grows around the right atrioventricular canal and merges with the tubercles of the atrioventricular canal cushions. Once fusion has taken place between the dextrodorsal conus ridge and the right tubercle of the ventral atrioventricular canal cushion, the communication between the right atrium and the common ventricle is closed The site of this earlier communication can be traced in the fully developed heart in the cephalic portion of the conus region of the left ventricle and just above the attachment of the septal cusp of the tricuspid valve in the right ventricle, it is known as the *atrio-*



Fig. 45.—Course of the crista supraventricularis and its bands in the right ventricle *A*, normal case, *B*, patent ductus arteriosus, *C*, sinus of Valsalva aneurysm, *D*, atrial septal defect. Structure and extent are the same in all With a left to right shunt with no rise in pressure, there is hypertrophy of the trabeculae (*B* and *D*), with a concurrent rise in pressure, there is hypertrophy of the actual wall muscles as well (*C*) *CSV*, crista supraventricularis, *I*, infundibulum; *PA*, pulmonary artery, *PB*, parietal band, *PM*, papillary muscle, *PSV*, pulmonary semilunar valve, *TV*, tricuspid valve, *SB*, septal band.

ventricular portion of the membranous part of the ventricular septum.

At the same time as the conus ridges sweep down toward the ventricular septum, the right tubercles of the atrioventricular canal cushions increase rapidly in size. Their enlargement is principally in the direction of the somewhat increased endocardial tissue masses of the crest of the ventricular septum. As early as the end of the second embryonic month, the endocardial tissue masses involved have grown to such an extent that they entirely bridge over the interventricular foramen. With the subsequent fusion, closure becomes complete. The end result is a fibroelastic connective tissue sheet, the *membranous part of the septum*. In the fully developed heart, it lies in the left ventricle in the conus region, slightly below the right and posterior semilunar valves (Fig. 46, and Fig. 81, p. 79). In the right ventricle, it lies immediately below the crista supraventricularis, behind the medial cusp of the tricuspid valve (Fig. 76, p. 74). It may sometimes herniate into the right ventricle (524) (Figs. 46 and 47).

If for some reason the conus ridges are laid down at an anomalous site, or have an abnormal course in the right ventricle, this chamber undergoes considerable modifications. If, as shown in Figures 48 and 49, the dextrodorsal conus ridge is absent or underdeveloped, a lengthy communication arises between the ventricles, and the right segment of the crista supraventricularis, as well as its parietal band, are wholly or partly lacking. If, on the other hand, this ridge is laid down at an anomalous site, or its growth in the normal direction is prevented for some reason, the result is the following. The interventricular foramen is not bridged, since the right segment of the crista supraventricularis grows in the anterior direction or even to the left, or backward to the left. The parietal band, which is not infrequently greatly foreshortened, therefore grows in the direction of the left segment of the anterior wall (Fig. 50) or toward the dorsal wall and the ventricular septum, then partly or wholly covering the septal band (Fig. 51). As a result, there is a considerable constriction of the outflow tract of the ventricle and a change in its mode of contraction. A concurrent developmental anomaly of the sinistroventral conus ridge is sometimes seen. This conus ridge may, however, be the only maldeveloped part, and then it usually grows in an abnormal direction. The left segment of the crista supraventricularis will then run in a more ventral direction, and the septal

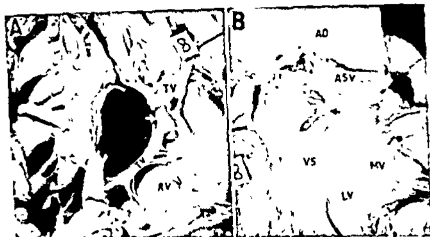


Fig. 46 —[illegible] ventricle Arr[illegible] and right ven[illegible]

into two halves, but they extend toward the common ventricle, at first in the direction of the ventricular septum. The subsequent growth of the two ridges nevertheless takes place in widely different directions. The *sinistroventral conus ridge* grows downward, forming the *septal band of the crista* the anterior ventricular wall, where it joins the trabeculae of this wall. Another part grows around the right atrioventricular canal and merges with the tubercles of the atrioventricular canal cushions Once fusion has taken place between the dextrodorsal conus ridge and the right tubercle



Fig. 45.—Course of the crista supraventricularis and its bands in the right ventricle *A*, normal case, *B*, patent ductus arteriosus, *C*, sinus of Valsalva aneurysm, *D*, atrial septal defect. Structure and extent are the same in all. With a left to right shunt with no rise in pressure, there is hypertrophy of the trabeculae (*B* and *D*), with a concurrent rise in pressure, there is hyper- [illegible] wall muscles as well (*C*), *CSV*, crista supraventricularis, *I*, infundibulum, [illegible] muscle, *PSV*, pulmonary semilunar valve,

supraventricularis, along the right side of the ventricular septum, toward the left caudal segment of the anterior ventricular wall, where it merges with its trabeculae (Figs. 36 and 45) The *dextrodorsal conus ridge* curves toward the *orifice of the right atrioventricular canal.* Part of it, the *parietal band of the crista supraventricularis*, continues toward the right caudal part of of the ventral atrioventricular canal cushion, the communication between the right atrium and the common ventricle is closed. The site of this earlier communication can be traced in the fully developed heart in the cephalic portion of the conus region of the left ventricle and just above the attachment of the septal cusp of the tricuspid valve in the right ventricle, it is known as the *atrio-*



... conus ridge, with communication be-
... (J E. 560108). The right ante-
... part of the septum, CSV, crista
... lunar valve.



Fig 50.—Tetralogy of Fallot A, the parietal band of the crista supraventricularis grows fo-ward and to the left, it is greatly foreshortened, causing marked stenosis of the ostium infu-dibuli B, anterior wall of the third ventricle lifted up AO, aorta, PA, pulmonary artery, P-parietal band, TV, tricuspid valve, VS, ventricular septum, VSD, ventricular septal defect

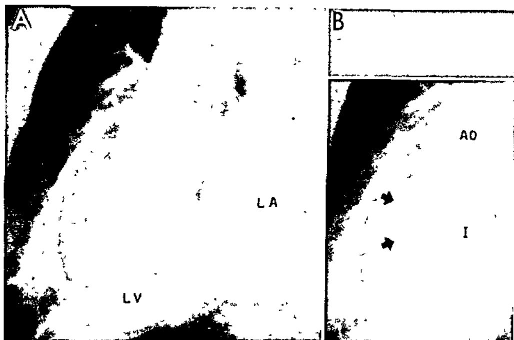


Fig. 47.—Herniation of the membranous part of the ventricular septum Girl, aged 8 (E S. 461008). Directly below the right anterior cusp of the aortic valve, the contrast medium is expelled into a fairly large pouch formation (arrows in *B*), which bulges into the right ventricle *AO*, aorta, *I*, infundibulum, *LA*, left atrium, *LV*, left ventricle.

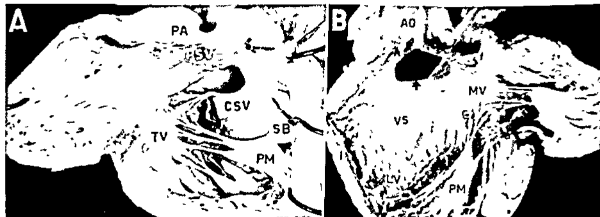


Fig. 48.—Absence of the dextrodorsal conus ridge, with wide communication between the conus region of the right and left ventricles The longitudinal course of the trabecular network along the diaphragmatic wall of the right ventricle can also be seen, running from the tricuspid orifice to the right corner of the ventricle *AO*, aorta, *CSV*, crista supraventricularis, *LV*, left ventricle, *MV*, mitral valve, *PA*, pulmonary artery, *PM*, papillary muscle, *PSV*, pulmonary semilunar valve, *SB*, septal band, *TV*, tricuspid valve, *VS*, ventricular septum



...ridge, with communication be-
(J E 560108). The right ante-
...art of the septum CSV, crista
...nar valve



Fig. 50.—Tetralogy of Fallot *A*, the parietal band of the crista supraventricularis grows forward and to the left, it is greatly foreshortened, causing marked stenosis of the ostium infundibuli. *B*, anterior wall of the third ventricle lifted up *AO*, aorta, *PA*, pulmonary artery, *PB*, parietal band, *TV*, tricuspid valve, *VS*, ventricular septum, *VSD*, ventricular septal defect

band is foreshortened and is fused with the anterior wall considerably more cephalad and farther to the right than normally (Fig. 52). This brings about a constriction of the outflow tract, particularly of the ostium infundibuli.

Ontogenetically, this constriction might be the result of deficient resorption of the so-called muscle ridge, which is found best developed in the reptilian heart (Fig 60, p. 59). Many features of the ontogenetic development of the heart are, in fact, reflected in the embryologic process in man Thus many of the developmental anomalies observed in the human heart are seen as the definitive stage in the lower groups of vertebrates. A comparison between the embryologic and ontogenetic development has been attempted in Figure 54. It is based on Streeter's division into horizons (632, 633, 634), each horizon corresponding to a period of about 48 hours.



Fig. 51.—Tetralogy of Fallot The parietal band is displaced dorsally and to the left, it lies ...) *A*, cusps of the tricuspid valve in place, ... the septal band The anterior cusp of the ... papillary muscles are small. *CSV*, crista ...l band, *PM*, papillary muscle, *RA*, right ...tricular septal defect

A definitive heart, derived from ventral vessel trunks, is found for the first time in animal series in Tunicata (647) However, as in Cyclostomata (suctorial fishes) and fishes, this heart pumps only venous blood It can be inferred from Figure 53 that in Elasmobranchii (cartilagenous fishes), for instance, the heart consists of sinus ve-



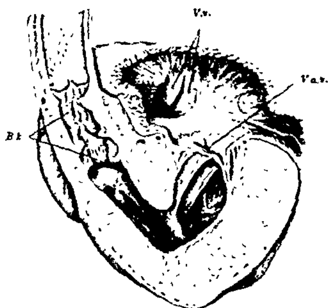


Fig 53.—Heart of Alopecias vulpes, opened from in front. *B.k.*, valves of bulbus; *V.a.v.*, valvulae atrioventriculares, *V.v.*, sinus valves (From Tandler, J: *Anatomie des Herzens* [Jena Gustav Fischer, 1913])

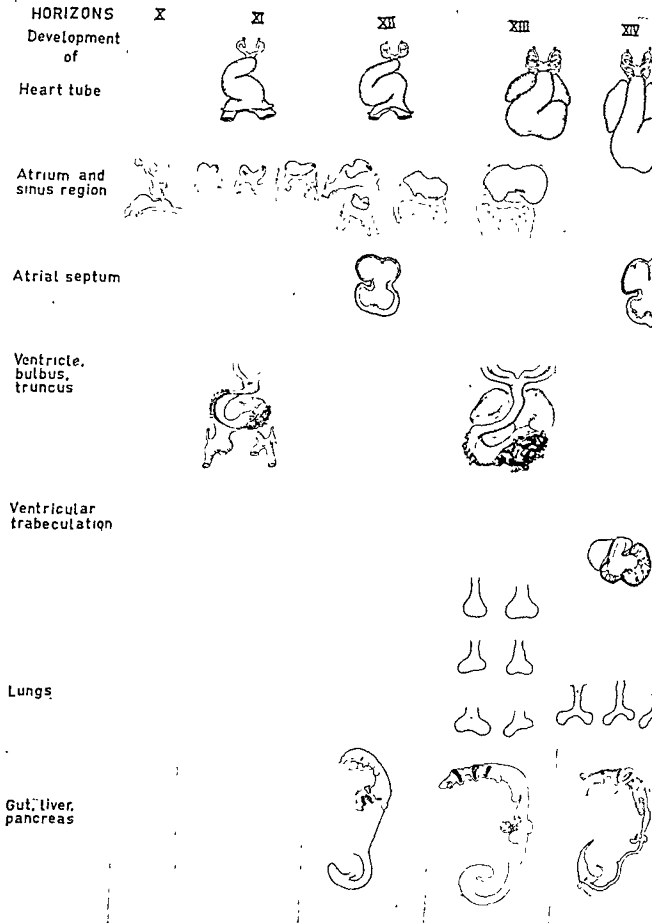
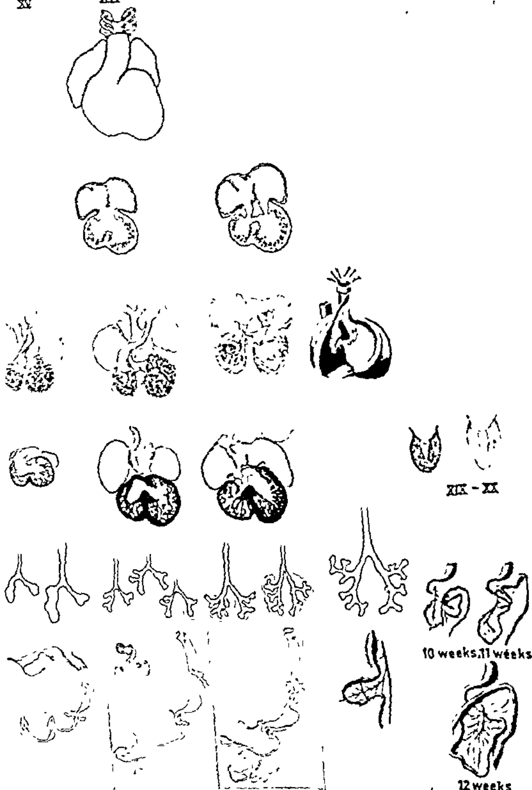


Fig. 54.—Comparison between embryologic and ontogenetic development of some organs (Asse…



leben *Handbuch der Anatomie des Menschen* [Jena Gustav Fischer, 1913]; Arey, L B,- *Developmental Anatomy* [Philadelphia W B Saunders Company, 1954]). (*Continued*)

HORIZONS X XI XII XIII XIV

Liver

Liver and pancreas

Stomach

Kidneys

Cyclostomata

Fishes

Amphibi

Heart

Lungs

XXI

Reptiles



Crocodiles









Fig 54 (*cont*)

nosus, atrium, ventricle, and bulbus cordis. The sinus venosus is a transverse, thin-walled sac, into which open the two ducts of Cuvier and the hepatic veins. The sinus venosus communicates with the symmetrical atrium through a transverse opening provided with two sinus valves (495). The atrium empties into the ventricle via a round or oval opening, the ostium atrioventriculare, in the left part of the ventricle. This opening has two valves.

From the right part of the ventricle is given off the bulbus cordis, which, accord-

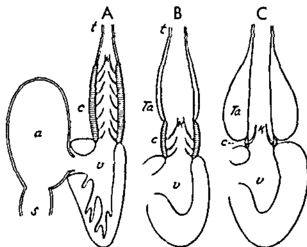


Fig. 55.—Schematic longitudinal section through the heart of different fishes A, Selachii, B, Amia, C, Teleostei: *a*, atrium, *c*, conus arteriosus (= bulbus cordis), *k*, valves, *s*, sinus venosus, *Ta*, truncus arteriosus, enlarged in C to bulbus arteriosus, *v*, ventricle (after Boas). (From Benninghoff, in Bolk, Goppert, Kallius, and Lubosch *Handbuch der vergleichenden Anatomie der Wirbeltiere* [Vienna. Urban & Schwarzenberg, 1953])

ing to Tandler (647), is that part of the outflow channel which has on its inner aspect the bulbar thickenings and their derivatives, the semilunar valves, and is covered externally by myocardium The bulbus cordis passes directly into the ventral aorta, its inner surface having rows of valves which prevent the blood from flowing back into the ventricle. This bulb is laid down in the embryo of all vertebrates, but regresses to a varying degree in ontogenesis (Fig. 55) It can be distinctly delimited only in Selachii (sharks), Ganoidei, Dipnoi (lungfish), and amphibians Regression takes place partly by incorporation of the bulbus region with the ventricle and partly by some of its distal part being replaced by the truncus. In the shark species Galeus, Mustelus, and Schyllidea, in Acipenser (sturgeon) and many other animals, the bulb has a form of valves, "Zungenklappen," which have been interpreted by Stohr (631) as "Taschenklappen" in a state of regression. They are situated below the true "Taschenklappen" (semilunar valves), have a broad attachment to the wall of the bulb, and protrude into the lumen like a lip with a rounded edge. Anatomically, the subaortic stenosis occurring in man (Figs 593, p. 655 and 597, p. 658) is reminiscent to some extent of these "Zungenklappen" (Fig. 56). Therefore it is conceivable that subaortic stenosis in man can be regarded as a developmental arrest, caused by failure of these valves to regress.

In the highest fishes, Dipnoi, in which the swim bladder serves as a kind of lung, a suggested division of the heart into an arterial and a venous part is found for the first time in the animal kingdom. Thus, the vein given off from the swim bladder empties directly into the atrium, its orifice being separated from the site of entry of the sinus venosus by a fold. In other fishes, the analogue of the pulmonary vein—the vein of the swim bladder—opens directly or indirectly into the sinus venosus According to Keith (378), this is a condition which may reappear as an abnormality in the human subject. Although in Dipnoi there is still intimate contact between the pulmonary vein and the sinus venosus—the former lying directly against the wall of the latter—the vein has a separate opening into the future left atrium, near the base of the left venous valve, in a manner almost identical with that shown in some human hearts with anomalous drainage of the pulmonary veins

In the amphibians, separation of the different chambers of the heart is still more advanced. The atrium is divided into a right and a left chamber by a septum, which grows from the posterosuperior wall of the

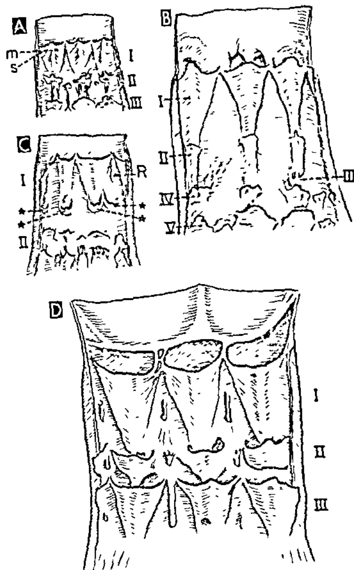


Fig 56 —*A*, conus of Mustelus vulg The thickened middle part (*m*) and the thin lateral

atrium (Fig. 57). The sinus venosus opens into the former chamber, and the pulmonary vein into the latter. The atrial septum has, however, a free, sickle-shaped margin, just above the ostium atrioventriculare common to the two atria. The features are somewhat similar to those of the developmental anomaly in man known as persistent ostium atrioventriculare commune. In amphibians, on the contrary, the ventricle and the bulbus cordis, as well as the truncus arteriosus, are not separated and lack any actual dividing walls. Structures are, however, present in the ventricle whose task is to prevent excessive mixing of the two blood flows entering it from the atria. They consist of a plentifully branched trabecular network, with the meshes arranged in such a way that those on the left mainly take up the blood from the left atrium, and those on the right, the blood from the right atrium.

If the development of the human heart is recalled, a somewhat similar arrangement of the ventricular musculature is found at a certain developmental stage (cf Figs. 57 and 58). In the human fetus, the endothelial heart tube is initially widely separated from the thick outer coat, the epimyocardium, which is still undifferentiated (19). The intervening space is filled with a fluid jelly, which serves the early pumping heart in a valvular capacity. The space is reduced as the jelly becomes transformed into the connective tissue of the endocardium (Fig. 58, *B*). The endocardial cushions and the bulbar thickenings are prominent because of an exaggeration and longer retention of this temporary condition The myocardial coat is, however, soon transformed, partly into a thick, spongy layer which largely fills the ventricular lumen in the form of a trabecular network (Fig 58, *C*) and partly into a thin, more compact cortical layer The spongy myocardial tissue is gradually condensed, especially peripherally, so that the superficial musculature becomes increasingly com-

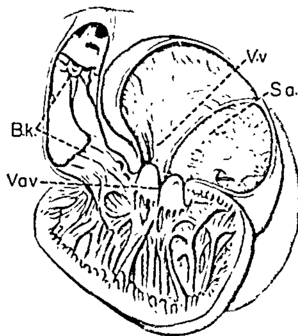


Fig 57.—Heart of Cryptobranchus japonicus. Atrium, ventricle, and bulbus opened from in front. *B.k*, *bulbus valves, S a*, *septum atriorum, which ends in a free margin just above the* ostium atrioventriculare, *V a v*, valvulae atrioventriculares, *V v*, sinus valve, to the right of it, a probe in the atrial orifice of the sinus venosus In the left atrium, the probe shows the *opening of the pulmonary vein. (From Tandler, J. Anatomie des Herzens [Jena Gustav Fischer,* 1913])

pact The trabeculae nearer to the lumen, on the contrary, retain an open arrangement for a long time (Fig. 58, *D*). It is this condition that is considered to be permanent in the lower vertebrates (19). The network in the left ventricle illustrated in Figure 59 may possibly represent incompletely resorbed muscular trabeculae. In mammals, the entire cardiac wall finally becomes compact. The irregular muscle bundles that persist beside the ventricular cavities make up the trabeculae carneae.

From the earliest stages, the primitive atrioventricular valve cusps are connected mains of the cusp may be present. According to Benninghoff (55), it lies apically to the borderline between atrial and ventricular septum, thus in the ventricle. A short portion of the ventricular septum therefore extends into the atrium. This is described by Benninghoff as follows (p. 521): "Die Klappenreste sitzen etwas apikalwarts von der Naht zwischen Verhof- und Kammerseptum, so dass ein kurzes Stuck des letzteren in den rechten Vorhof ragt. Es ware das ein muskuloses Septum atrioventriculare." The description is highly reminiscent of that generally given of the

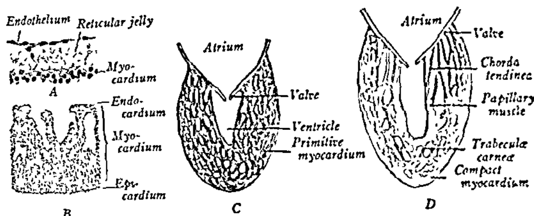


Fig 58 —Differentiation of the human ventricular wall and the atrioventricular valves A and B [illegible] 7 mm (× 55), respectively. C and D, diagram- [illegible]). (From Arey, L. B : Developmental Anatomy [illegible]

with the network of muscular trabeculae which traverse what is to become the cavity of the ventricles (Fig 58, C). According to Odgers (512), in a certain stage the valve cusps consist almost exclusively of muscular tissue This regresses, however, and is replaced by collagen tissue, the cusps then assuming their definitive appearance of a thin valve This regression may fail to take place or may be deficient This applies in the crocodile, Sarcorhamphus (a species of vulture), Occydromus (Maori hen), Corvus mandula (a species of crow), and the turkey In these animals, the medial (septal) cusp of the right atrioventricular orifice is wholly or partly converted into an endocardial ridge, on which a small, free remalformation of the tricuspid valve in man known as Ebstein's disease. This may possibly confirm the assumption of Baker *et al* (36) and Odgers (512) that this anomaly arises during early embryonic development.

Cardiac malformations in combination with asplenia have sometimes been observed in man (356, 448, 515). It is known from the investigations of Arey (18), Broman (102), and Ivemark (356), among others, that the human splenic primordium can be definitely traced as early as in the XIVth to XVIIth horizon (Broman, XIIth horizon) In severe developmental disturbance of the spleen (asplenia), damage to the splenic primordium is considered to



Fig. 59.—Left ventricle crisscrossed by a network of trabeculae, reminiscent of those in Figures 57 and 58 Boy, aged 5 months (S G-V 530830). An atrial septal defect and infundibular stenosis are also present (see Fig 399) In *B*, the network has been partly cut through, and in *C*, completely cut through, to display the interior of the ventricle.

have occurred at the aforementioned early stage. In coincident asplenia and cardiac malformation, there is good reason to assume that the damage has involved the primordia of both organs and that this has occurred at about the same time in the two cases, at a stage corresponding to the XIVth to be considered typical of this developmental syndrome (448). Thus, on broad lines, the picture is in agreement with the description of the human embryonic heart in the XIVth to XVIIth horizon. From the ontogenetic point of view, it might be said that the development of the human heart

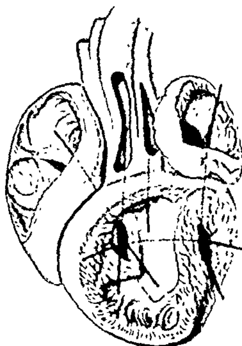


Fig. 60 —Heart of Varranus niloticus. Both atria, both ventricles, the pulmonary artery, and right aorta are opened from in front. The probe introduced through the left atrium emerges, [illegible] behind the muscle ridge into the right aorta. The probe [illegible]nning horizontally goes from the an- [illegible] incompletely divided posterior ven- [illegible] er, J.: *Anatomie des Herzens* (Jena. [illegible]

to XVIIth horizon At this stage of development, division of the heart into four separate chambers is not complete, the atrioventricular cushions have not yet fused, and the conotruncus is incompletely divided The cardiac malformations observed in man with coincident asplenia are generally multiple and are characterized by faulty septation of the [illegible]

has been wholly or partially arrested at the amphibian to reptile stage.

In this connection, it should perhaps be mentioned that the cardiac anomalies in man found in combination with less-advanced splenic malformation (multiple spleen) are of an entirely different nature and of less serious character. This is compatible with the assumption that damage has occurred at a later, less sensitive stage in development of the heart and spleen

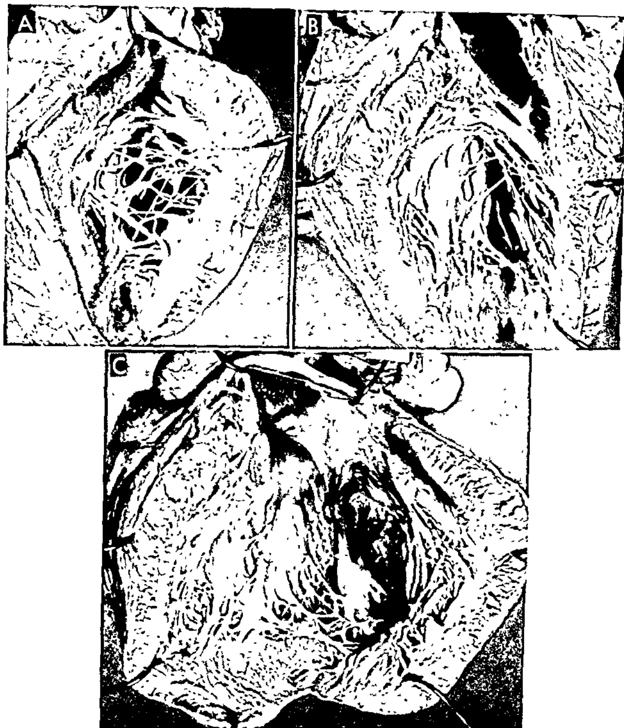


Fig. 59.—Left ventricle crisscrossed by a network of trabeculae, reminiscent of those in Figures 57 and 58. Boy, aged 5 months (S.G-V. 530830). An atrial septal defect and infundibular stenosis are also present (see Fig 399) In *B*, the network has been partly cut through, and in *C*, completely cut through, to display the interior of the ventricle.

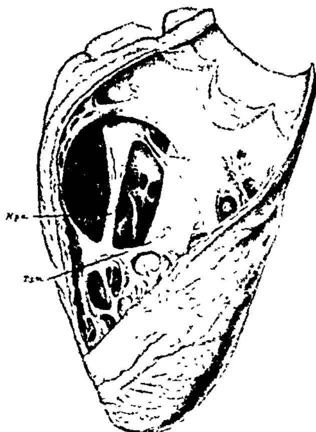
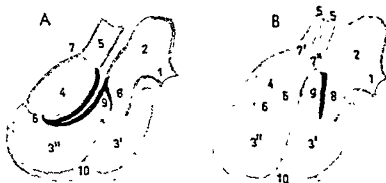


Fig. 61.—Human right ventricle, opened from in front. The infundibulum is separated from the ostium venosum by the crista supraventricularis There is distinct trabecula septomarginalis (T s m), which is intimately joined to the anterior papillary muscle (M p a.). (From Tandler, J *Anatomie des Herzens* [Jena Gustav Fischer, 1913])



B, diagram-
3'', left and
7, 7' and 7''
junction, 9, anterior cusp of mitral valve, 10, base of
interventricular septum (Redrawn from Keith, A *Human Embryology and Morphology* [Lon-
don Edward Arnold & Co , 1948])

In reptiles, the separation of the heart into different chambers has proceeded further (Fig. 60). The atrial septum has grown down into the opening between atrium and ventricle, and the truncus arteriosus has become divided into three trunks: the pulmonary artery and the right and left aorta. As a result, the atria—which are completely separated from each other—empty into the ventricle through their own atrioventricular orifices. Division of the ventricle has also started to take place. Development is, however, advanced to a varying degree in the different species, it is least advanced in the lizard and tortoise and most advanced in the crocodile. In the first-mentioned, the ventricle is incompletely divided by a septum—the muscle ridge (Fig. 60)—into a smaller ventral chamber lying to the right, cavum ventrale, which passes anteriorly into the pulmonary artery, and a larger dorsal chamber, lying to the left, cavum dorsale Both atrioventricular ostia open into the latter, and both aortae are given off from it The muscle ridge takes its origin from the apex and left part of the anterior ventricular wall and runs obliquely backward to the right. It has a free margin, concave upward, which is not attached to the heart wall. A communication between cavum dorsale and ventrale therefore persists (Fig 60). The dorsal chamber has a suggested division into two lateral halves, the right and left cavum ventriculi dorsale, by a sagittally placed elevation on the posterior wall of the ventricle, the trabecular ridge. This ridge bulges forward in the direction of the ventral part of the muscle ridge. The blood from the atria empties through the respective orifices into the two aforementioned parts of the cavum dorsale The arterial blood flows into the left part, and the venous blood into the right, whence it passes over the free margin of the muscle ridge into the anterior right ventricle and is then pumped into the pulmonary circulation. The arterial blood flowing into the left part is, however, mixed to some extent with the venous blood before it is pumped into the aortae.

In the crocodile, division of the ventricle into two separate halves is more advanced. The sagittal trabecular ridge is more strongly developed; it is continuous both with the ventral part of the muscle ridge and with an endocardial septum growing out from the wall dividing the aortic orifices. In this animal, the part of the muscle ridge to the right of the site of insertion of the trabecular ridge has markedly regressed, so that the right cavum dorsale and the cavum ventrale have become a single chamber.

According to Tandler (647), the right cavum dorsale lying behind the muscle ridge corresponds to the inflow tract of the right ventricle in mammals, and the cavum ventrale lying in front of the muscle ridge, to its outflow tract. The rudimentary right part of the muscle ridge in the crocodile has its analogue in mammals in the trabecula septomarginalis (Fig. 61), described by King (387) and Retzer (555) as the moderator band.

When resorption of the right part of the muscle ridge fails to occur or is defective, the prerequisite exists for development of a constriction of the channel joining the inflow and outflow tract of the right ventricle. This is probably the factor responsible for the infundibular stenosis illustrated in Figures *216* and *399b*. Such an *assumption* seems plausible, even against the background of Keith's (378) view that the fourth chamber of the human heart—the bulbus cordis (*4* in Fig. 62, *A*)—becomes submerged in the ventricles, principally in the right ventricle (Fig. 62, *B*), just as the sinus venosus becomes included in the atria For, by atrophy of the muscle wall (represented by a heavy black line in Fig. 62, *A*) between the bulbus cordis and the right ventricle in the second fetal month, the cavity of the bulbus is thrown into that of the ventricle, and the atrioventricular and aortic orifices are brought side by side.

In the crocodile, the events related above produce a complete ventricular septum for the first time in the vertebrate series. Di*rectly below the aortic orifices this septum*

branchial region takes place, and a *common dorsal aorta* is formed

Here, as at other sites in the body, the degenerative processes follow close on the new formations. Even before the last pair of aortic arches has developed, the first two and the fourth aortic arch on the left side, and between the third aortic arch and the common aorta on the right side.

Of the then remaining branchial arches, the left third arch becomes the *definitive aortic arch*, whereas the ventral part of the



Fig 64 —Some of the commonest vascular anomalies of the aorta and brachiocephalic arteries *A*, right aortic arch with right descending aorta, *B*, left aortic arch with anomalous right subclavian artery, *C*, right aortic arch with descending aorta and aortic diverticulum with anomalous left subclavian artery. *D*, double aortic arch, *F*, coarctation of the aorta, fetal type, *F*, absence of both common carotid arteries *I–VI*, first to sixth branchial arch arteries, *AscAo*,

have disappeared entirely In a 4-week-old fetus, only the first four pairs of aortic arches are developed, as may be inferred from Figure 63 Two weeks later, when the sixth pair is fully developed, the first two pairs have already degenerated. Simultaneously, there is disappearance of the parts of the dorsal aortae lying between the third sixth forms the *pulmonary arteries*, and its dorsal part the *ductus arteriosus* (Fig. 63). Vascular anomalies in this region are extremely common. Many of them are of no practical consequence, but from a differential diagnostic viewpoint it is important to be familiar with them. The commonest malformations are shown in Figure 64

is, however, perforated by a small hole, the foramen of Panizza. Consequently, the two circuits must be regarded as incompletely separated from each other. Separation only becomes complete in birds and mammals. this way, the aortae grow headward, but curve dorsally at the level of the pharynx and then follow the whole primitive spine caudally as the *dorsal aortae* The communications present between the ventral

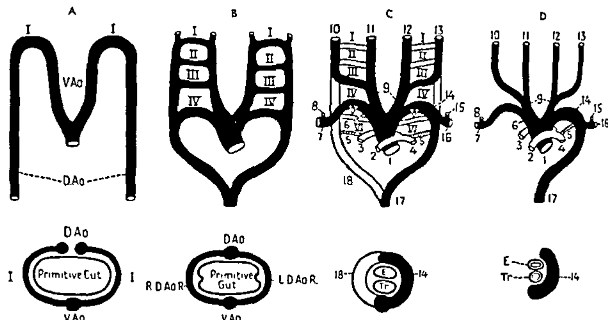


Fig. 63.—Outline of fetal development of the arterial trunk and brachiocephalic arteries (adapted from several sources) *Upper row*, frontal view of the primitive vascular bed. *Lower row*, view from above (from Eek, in Mannheimer, E.: *Morbus Caeruleus* [Basel S Karger, 1949]). *A*, 2–3 weeks. *I*, first branchial arch artery, *DAo* and *VAo*, dorsal and ventral aortae *B*, 4 weeks. *I–IV*, first to fourth branchial arch arteries, *DAo* and *VAo*, dorsal and ventral aortae, *LDAoR* and *RDAoR*, left and right dorsal aortic roots. *C*, 6 weeks, and *D*, at birth *I–VI*, first to sixth branchial arch arteries, *1*, ascending aorta, *2*, pulmonar[illegible] monary arter[illegible] vertebral arte[illegible] and left exter[illegible] tebral artery, *16*, left subclavian artery, *17*, left descending aorta, *18*, right dorsal aortic root, *E*, esophagus, *Tr*, trachea

C. DEVELOPMENT OF THE LARGE ARTERIES

The development of the arteries takes place, on broad lines, parallel to that of the heart. Already in the second embryonic week, small cellular strands of mesodermal origin can be observed cephalad to the primordium of the heart. They become hollowed out and give rise to the *ventral aortae* (91). Where main blood vessels are about to be laid down, a meshwork of these small channels is found. Gradually, some of these primitive channels are enlarged and straightened to form the main blood vessels, and their walls become thickened In and dorsal aortae, which are at the level of the mandibular arch, are known as the *first aortic (branchial) arches* (Fig. 63). During the subsequent period, five additional pairs of aortic arches are developed between the ventral and the dorsal aortae, although the fifth pair is usually rudimentary in mammals. Each of these arches runs in the corresponding visceral arch in a direction caudal to the mandible

Concurrently with the fusion of the two cardiac primordia into a single tube, the proximal parts of the ventral aortae become fused to form the ascending aorta. In the fourth embryonic week, a similar fusion of the *dorsal aortae* caudal to the

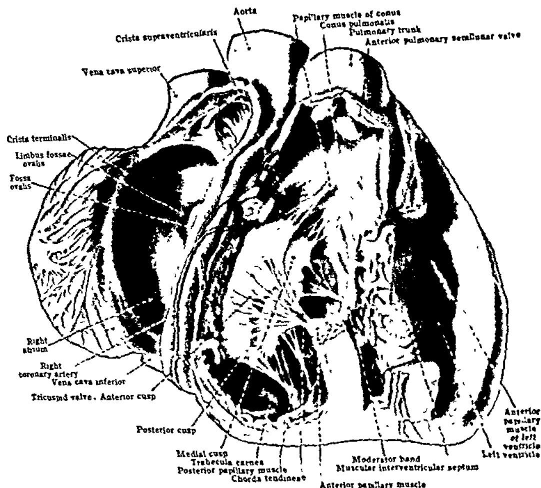


Fig 65.—Ventral view of the heart with walls of the right atrium and ventricle opened to show their internal configuration The dissection was planned to show the relations of an unusually well developed moderator band Position of the foramen ovale is somewhat more cephalic than usual (From *Morris Human Anatomy* [10th ed , New York Blakiston Company, 1942])

2 Roentgenologic Anatomy and Physiology of the Heart

THE OBJECT of the present chapter is to give an account of such anatomic details and physiologic features as we consider to be of particular importance for the angiocardiographic interpretation

The *right atrium* consists for the greater part of the atrium of the embryonic heart. Only a small part is formed from the incorporated *sinus venosus*. The borderline between them is marked by a powerful muscular ridge, the *crista terminalis* (Figs. 65 and 66) A muscular network (Fig. 67), extending from the crista to the *annulus fibrosus*, is mainly responsible for the thickness of the wall. In the grooves between the muscular ridges, the wall is, on the contrary, extremely thin. The structure of the network gives reason to suppose that the movement of the plane of the tricuspid valve toward the posterior atrial wall during atrial systole is brought about chiefly by its coarser muscle fibres, which run fairly parallel from the crista terminalis toward the annulus fibrosus. The superior and the inferior parts of the atrium are then brought closer together, whereas the position of the posterior wall is practically unchanged. This is presumably due to the fact that it is anchored by the two venae cavae (Figs. 68 and 69).

A similar muscular network spreads from the crista terminalis around the whole inner wall of the *auricular appendage*, as far as its apex (Fig. 65). It may be inferred from Figure 69 that the appendage extends far medially, usually as far as the aortic root and the conus region of the right ventricle. Consequently, at angiocardiography performed intravenously or by injection directly into the right atrium (Fig. 70), the evaluation of the conus and the pulmonary orifice is hampered by the fact that they are partially overlapped by the contrast-filled auricular appendage. During atrial systole, the appendage moves to the right and becomes contracted at the same time. This concentric contraction of the appendage starts in its apex and continues as a squeezing movement toward the base (Fig. 69)

With increased diastolic filling of the atrium, the aforementioned muscular network hypertrophies, whereas the rest of the wall does not become thickened, or only inappreciably (Fig. 67, *E*)

As shown in Figures 65, 66, and 68, the inner surface of the sinus venosus is completely smooth. The *superior vena cava* and the *inferior vena cava* open into its superior and inferior part, respectively. It may also be inferred from Figure 69, *G–J*, that these two veins form a slight angle, which becomes somewhat widened during atrial systole. At angiocardiography, the *valve of the inferior vena cava* can sometimes be identified at the mouth of this vein (Figs. 69, *I–J*, and 73–75) The coronary sinus opens at about this level and slightly to the left (Figs 73, 74, and 76). On the posterior

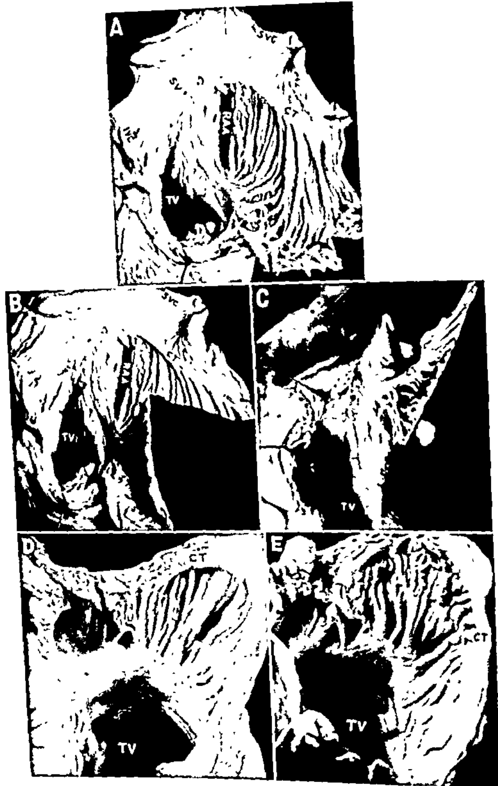


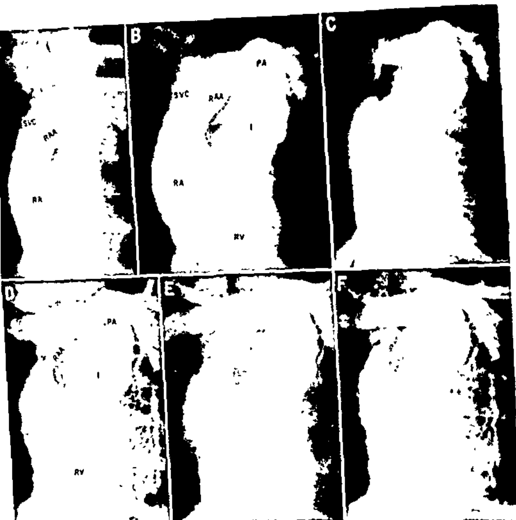
Fig 67 (legend on facing page)



Fig. 66.—Right atrium *A*, sinus venosus cut and laid open, *B–D*, crista terminalis (arrow in *A*)—from in front (*B*), severed at the inferior vena cava (*C*) and laid open to show the anterior atrial wall (*D*). *CT*, crista terminalis, *IVC*, inferior vena cava, *RA*, right atrium; *SV*, sinus venosus, *SVC*, superior vena cava

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Fig. 67.—Trabecular network of the right atrium Trabeculae run radially from the crista ter- [illegible] C, the thinness of the wall between [illegible] ied, the trabeculae lie close together [illegible] in the network are widened (*B* and *C*). *D*, primary pulmonary hypertension, *E*, atrial septal defect. In both, there is considerable hypertrophy of trabeculae *CT*, crista terminalis, *IVC* and *SVC*, inferior and superior venae cavae, *RAA*, right auricular appendage, *SV*, sinus venosus, *TV*, tricuspid valve ⟶



[illegible]

vena cava (*continued*)

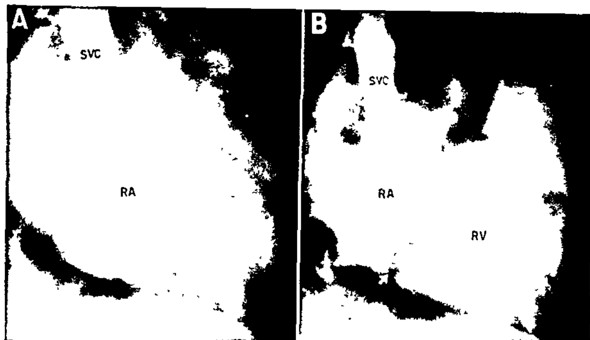


Fig. 68.—Right atrium, patent ductus arteriosus (M.N 420410). *A*, late diastole, *B*, late systole. During both phases, position of the dorsal wall of the atrium is practically unchanged, whereas the atrioventricular border moves far dorsally. In *B*, arrow points to the annulus fibrosus. *RA*, right atrium; *RV*, right ventricle, *SVC*, superior vena cava.



Fig. 69 (*cont*) —*L–Q*, conditions during filling and contraction in left atrium and ventricle. Arrow in N points to atrioventricular plane

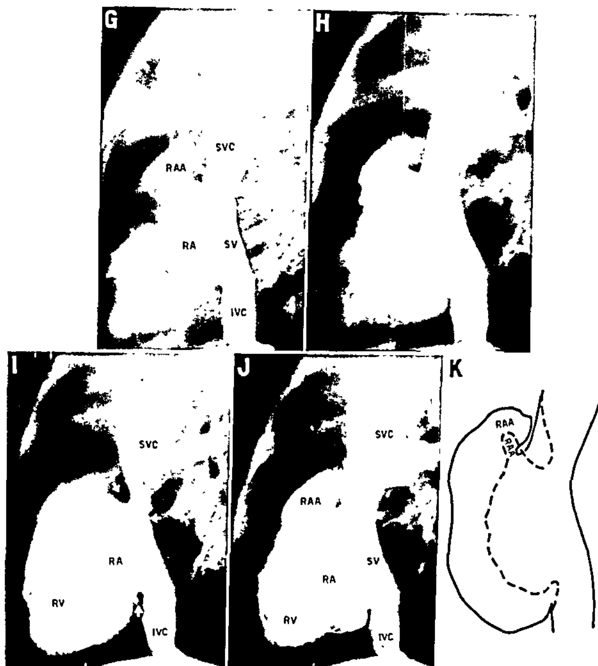


Fig. 69 (*cont*) —G–J, during atrial systole, the atrioventricular border is shifted dorsally, while the dorsal wall of the atrium remains in the same position The crista terminalis presses into the lumen like a membrane (*I*, lower arrow) Sphincter mechanism of the venae cavae is clearly visible. *K*, collective picture of appearance of atrium in late diastole (solid line) and late systole (broken line) (*Continued*)



Fig 73.—Right atrium *A*, valves of the vena cava and the coronary sinus (the latter perforated), *B*, large valve of the vena cava, extending toward the superior vena cava *FO*, foramen ovale, *IVC*, inferior vena cava, *TV*, tricuspid valve; *VCS*, valve of coronary sinus, *VVC*, valve of vena cava.

Ligamentum arteriosum
Left pulmonary artery
Aorta
Probe in transverse sinus
Right pulmonary artery
Right auricle
Left pulmonary veins
Vena cava superior
Left atrium
Right ventricle
Foramina venarum minimarum
Crista terminalis
Right pulmonary veins
Interatrial sulcus
Left ventricle
Limbus fossae ovalis
Right atrium
Fossa ovalis
Tricuspid valve:
Orifice of coronary sinus
Anterior cusp
Valvula sinus coronarii
Medial cusp
Valvula venae cavae inf
Posterior cusp
Vena cava inferior
Musculus pectinatus
Right coronary artery
Small cardiac vein

Fig 74 —[illegible]
its internal cc [illegible]
pany, 1942])

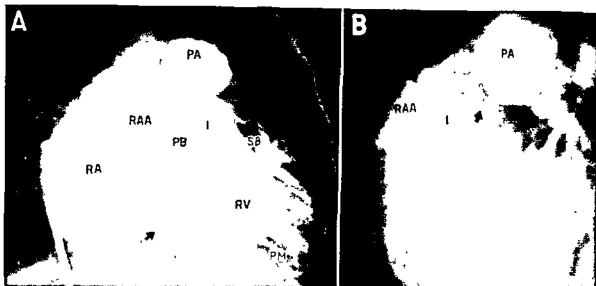


Fig. 70.—Valvular pulmonary stenosis. Girl, aged 2 (M.S. 510423). The right atrium is large and the appendage extends as far as the infundibulum. In lateral projection (*B*) it overlaps the infundibulum and valvular region, increasing the difficulty in evaluating this area Arrow in *A* points to the atrioventricular border, arrow in *B*, to the fused cusps *I*, infundibulum, *PB*, parietal band, *PM*, papillary muscle, *RA*, right atrium; *RAA*, right auricular appendage; *RV*, right ventricle, *SB*, septal band, *PA*, pulmonary artery.



Fig. 71 (*left*) —Tricuspid orifice seen from above The medical cusp (*TV*) bulges into the orifice

Fig. 72 (*right*).—Tricuspid and mitral orifices seen from above Atrial wall is cut away as far as the atrioventricular border The annuli fibrosi (solid black line) are obliquely placed, their highest point is at the level of the aortic root and their lowest, posteriorly to the right and to the left, respectively. *LA* and *RA*, left and right atria

wall of the atrium is seen a slight impression (Fig 69, *G–H*), caused partly by the right inferior pulmonary vein (Figs. 77 and 78) and sometimes by a local thickening of the wall as well.

In both animals and man, the muscles of the atrial wall extend onto the extremities of the superior and inferior venae cavae

tion of the right side, and that the central ray be directed 20–30° cranially. The anterior cusp of the tricuspid valve is usually smaller than the others. Since the ventricular septum bulges toward the right ventricle, the medial cusp of this valve deviates to the right. The channel formed by the cusps is therefore directed toward the left

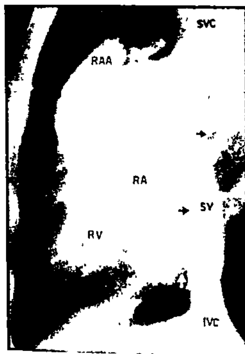




Fig 77 (left)

ca
ct

(548, 562) This musculature, which continues farther onto the superior than onto the inferior vena cava, surrounds the vessels like a cuff The muscular rings contract strongly during atrial systole (120, 325, 395) and probably act as sphincters (Figs 69 and 79).

Optimal visualization of the tricuspid orifice requires that the patient be in the oblique position, with a 45° anterior rota-

caudal corner of the right ventricle (Fig. 71)

The *left atrium* differs considerably from the right, with respect both to its structure and to its mode of contraction The wall is thicker and consists of muscles with a circular arrangement, only the appendage is trabeculated. The parts of the veins incorporated with the atrium occupy a consider-

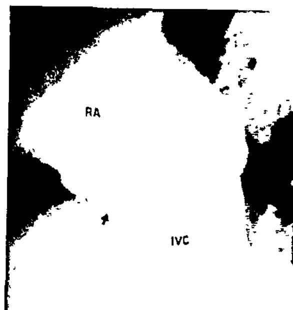


Fig. 75.—Valve of the vena cava. *IVC*, inferior vena cava, *RA*, right atrium

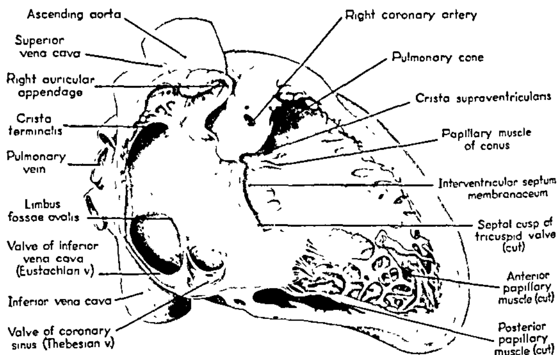


Fig. 76.—Right side of heart opened in a plane approximately parallel to the septa to show
tricuspid
the inter-
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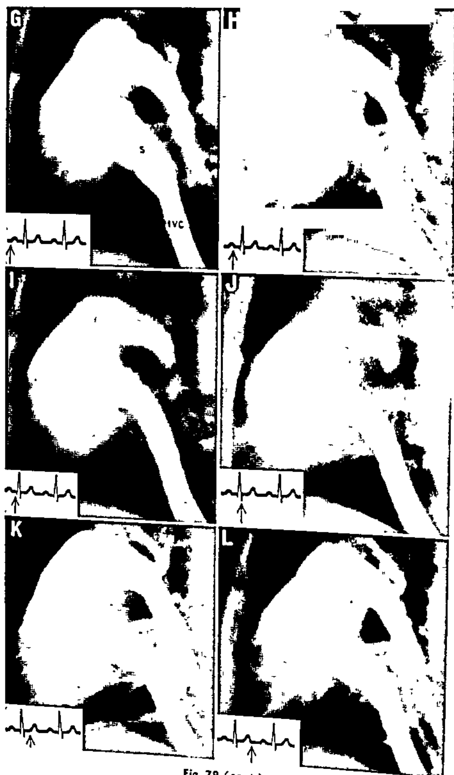


Fig 79 (*cont.*)

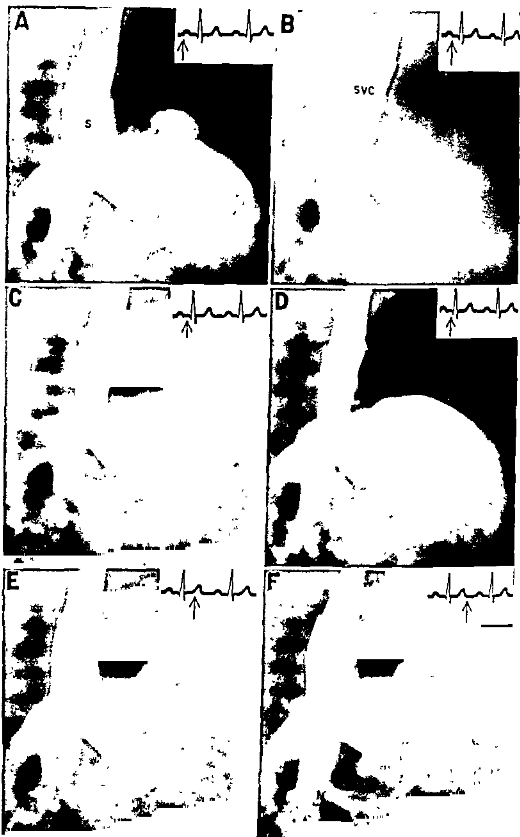


Fig 79 —Variations in the lumen in the extremities of the superior (A–F) and inferior (G–L)

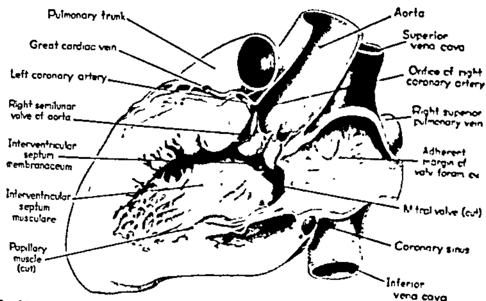


Fig 81.—Left side of heart opened in plane

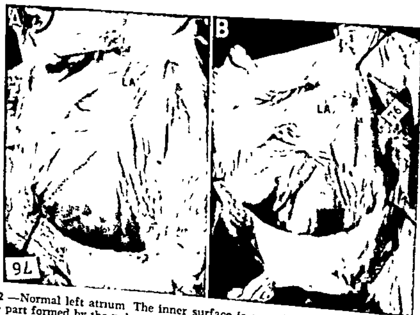


Fig 82 —Normal left atrium The inner surface is smooth, with no distinct transition between the part formed by the pulmonary veins and the rest of the atrium The wall is considerably thicker than that of the right atrium (cf Fig 67, p 67) *LA*, left atrium

ably larger area than they do in the right atrium. This is apparent from Figure 80, in which no pulmonary veins have taken part in formation of the atrial wall. All these *veins drain through the left superior vena* cava. The rest of the atrium consists mainly of an appendage of fairly normal size. This is in good agreement with Keith's (378) view that, to all intents and purposes, the primitive left atrium forms the appendage only.

Under normal conditions, the part formed by the veins passes without any marked transition into the remainder of the atrium (Figs 81 and 82). According to Keith (378), this venous segment is demarcated from the rest of the atrium by a prominent muscular fasciculus — the *taenia terminalis sinister* The superior pulmonary veins usually open laterally and anteriorly to the inferior pulmonary veins (Fig. 3a, p. 4). The two groups are generally separated by the main bronchus, so that the superior vein passes in front of it and the inferior behind it (Fig. 83). On the right side, the superior pulmonary vein runs in approximately the same plane as the main branch of the pulmonary artery or slightly anterior to it. On the left side, the superior pulmonary vein runs partly in front of this artery.



Fig. 80.—Small left atrium in complete anomalous venous return to the left superior vena cava (R J 520127) *LA*, left atrium, *LAA*, left auricular appendage

The annulus fibrosus is demonstrated in Figures 71 and 72. In order for the valvular plane to be depicted with the least possible distortion, the patient must be rotated 45° and the central rays directed 20–30° cranially. The anterior cusp of the mitral valve is generally larger than the posterior and, as a rule, is also somewhat thicker. The posterior cusp is sometimes partly divided into two portions, which are separated by a group of chordae tendineae.

Probably because of the special arrangement of the muscles, the whole atrium contracts concentrically (688), as may be inferred from Figures 69, *L–Q*, and 84. During atrial systole, the atrioventricular border is drawn toward the veins, whereas the posterior wall is fairly firmly anchored by the pulmonary veins According to Arvidsson and Odman (23), the atrium is almost completely emptied in atrial systole, this could be confirmed in our series.

The *left auricular appendage* extends toward the conus region of the right ventricle, between the pulmonary artery and the left ventricle. Its mode of contraction is strongly reminiscent of that of the right (Fig. 85).

As on the right side, the wall muscles continue onto the veins and encircle them These muscular rings contract during atrial



Fig 84 —Mode of con... with the atrio...

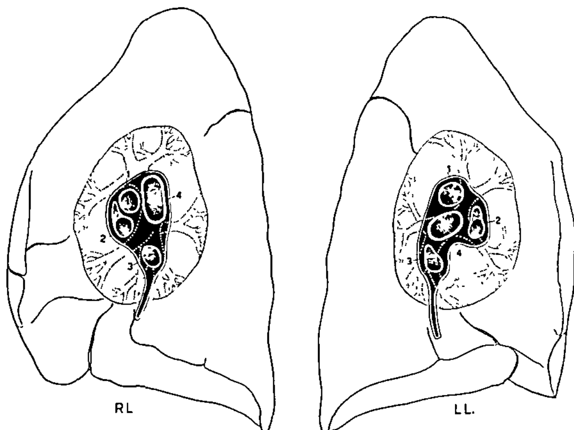


Fig. 83.—Position of the pulmonary veins in relation to the bronchi and pulmonary artery in the hilar region. *1*, pulmonary artery, *2* and *3*, superior and inferior pulmonary veins, *4*, main bronchus, *RL* and *LL*, right and left lungs.

systole. They probably act as sphincters (548) (Fig. 86).

THE VENTRICLES.—The *right ventricle* differs in many respects from the left; this applies to both its structure and its function. As seen in Figures 65 and 69, *A–F*, it may be compared to a pyramid, with the ... basal corner and the ... The ... *en-*

seldom be identified. During ventricular diastole, only the orifice can as a rule be visualized (Fig. 88, *B*), and the annulus surrounding it appears as a filling defect in the end-diastolic phase (Fig. 68, *B*). The contrast medium assembled in the pocket between the valvular cusps and the ventricular wall is occasionally seen as a ring surrounding the cusps (Fig. 89).

The part of the right ventricle at the level

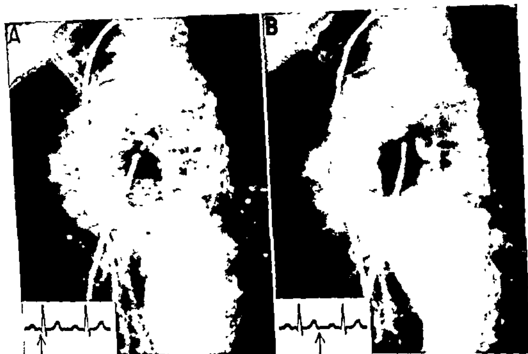


Fig. 86.—Changes in lumen in the proximal part of the pulmonary veins (arrow). *A*, strong circular contraction in atrial systole, *B*, relaxation during ventricular systole.

tricularis into two parts differing both in appearance and in function (42, 525, 559, 648) The *sinus* or *inflow tract* caudal to the crista supraventricularis is strongly trabeculated (Figs 65 and 87), and *papillary muscles* arise from its walls At angiocardiography, the anterior papillary muscle can usually be visualized, and sometimes the posterior as well A definite change in their shape occurs during the cardiac cycle (Fig 437, p 470)

A bulging valvular plane may be observed during systole (Fig 88, C), but the individual cusps of the tricuspid valve can

of the crista and above it, the *infundibulum*, *conus arteriosus* or *outflow tract*, has, in contrast, a practically smooth wall and no trabeculae in its posterior part. The infundibular opening, the *ostium infundibuli*, is sometimes slightly narrowed in the anteroposterior direction, owing to bulging of the crista supraventricularis from behind (Figs 88 and 90). The *bands* issuing from the crista, the *parietal* on the right side and the *septal* on the left, run like two ribbons caudally and forward and merge with the trabeculae of the inflow tract (Fig. 90) They are seen on the roentgenogram



Fig. 85.—Mode of contraction of the left auricular appendage Contraction starts in the apex and continues toward the base. A and B, early, and C, late atrial systole. *LAA*, left auricular appendage.



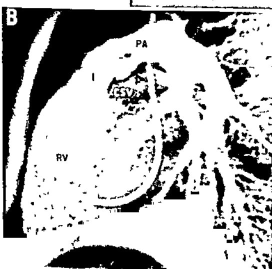




Fig. 88 —Valvular pulmonary stenosis. Cusps of the [illegible]

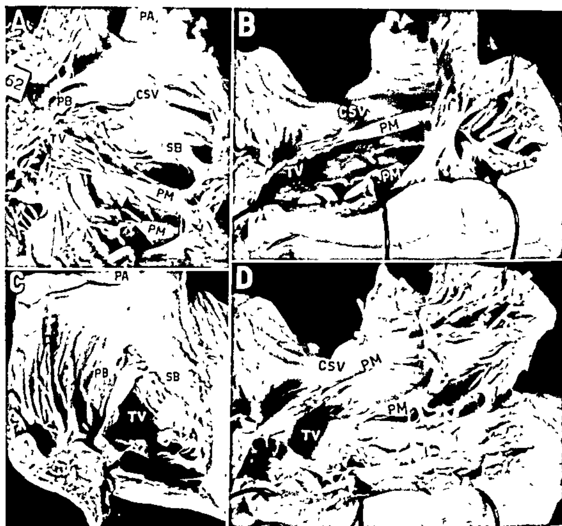


Fig. 87.—Right ventricle. *A*, ventral wall reflected; *B*, from the diaphragmatic surface, *C*, [illegible] corner reflected to show course of the [illegible] aventricularis, *PA*, pulmonary artery, [illegible] *TV*, tricuspid valve.

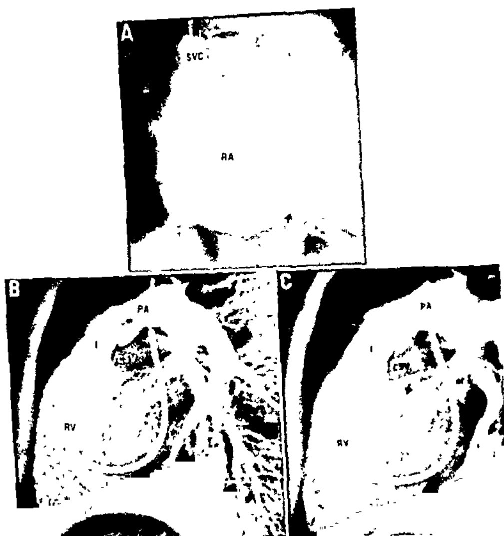


Fig 88.—Valvular pulmonary stenosis. Cusps of the tricuspid valve. Individual cusps are usually difficult to identify. The valvular plane can, however, often be visualized; its appearance varies with the cardiac phase *A*, filling of the right atrium during late ventricular systole (arrow points to the valvular plane), *B* and *C*, injection into the right ventricle, *B* in late diastole, *C* in early systole CSV, crista supraventricularis, *I*, infundibulum, *PA*, pulmonary artery, *RA*, right atrium, *RV*, right ventricle, *SVC*, superior vena cava.

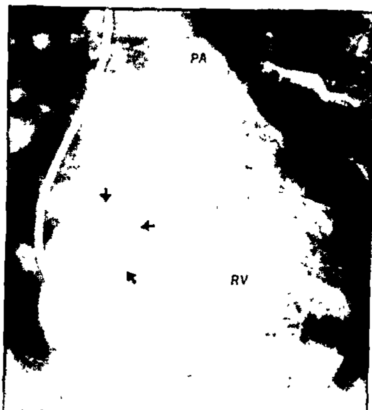


Fig. 89.—During diastole, contrast medium collects between cusps of the tricuspid valve and ventricular wall. The cusps may then appear as a ring-shaped filling defect (arrows) *PA*, pulmonary artery, *RV*, right ventricle

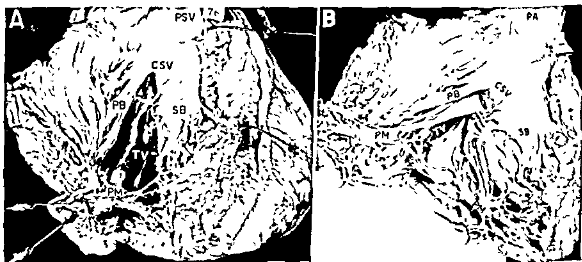


Fig. 90.—Normal case The right ventricle is cut open along the septal border The septal band, which is thicker and more compact, runs along the septum toward the apex and joins the trabecular network The parietal band, which is thinner and segmented, runs along the right [illegible] part, where it is incorporated with the trabeculae. Position of the [illegible] -n- [illegible] ry artery, *PB*, parietal band, *PM*, papillary muscles, *PSV*, pulmonary semilunar valve, *SB*, septal band, *TV*, tricuspid valve

as two powerful ridges of variable thickness (Fig 201, p. 210). The parietal band gives off a fasciculus which partly surrounds the *ostium venosum*. On the inner aspect of the anterior and right wall is found a longitudinal trabecular network which extends from the region of the semilunar valves toward the base of the ventricle (Figs. 87 and 90). A similar trabecular system runs from the tricuspid area toward the left caudal corner of the ventricle (F... 87)...

segment of the ventricle (Fig 87). A thick

trabeculae, which hypertrophy. Hypertrophy of the actual ventricular wall takes place when the ventricle is forced to work under an increase of pressure (Fig. 93).

The difference between the thickness of the walls of the two ventricles is considerably less in the newborn infant than later in life. This is shown, for example, by our study of nine full-term fetuses without cardiac malformations Even at this time, however, the wall of the left ventricle is thicker than that of the right. It is true that the wall of the latter is apparently thick, but this must be ascribed more to a strongly

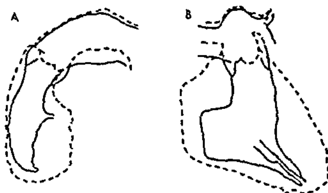


Fig 91.—Mode of contraction of the right ventricle Broken line, late diastole, unbroken line, midlate systole

muscular trabecula, the *moderator band* (42, 657), sometimes issues from the septal wall, it is attached to the base of the anterior papillary muscle (Fig 65).

During ventricular systole, the crista supraventricularis and its two bands are seen to draw the floor of the ventricle like a piston toward the outflow tract, while the ventricular wall is contracted and the base of the ventricle is drawn toward the apex and the conus (Fig 91). Since the heart is fixed to the surface of the diaphragm, the former movement nevertheless causes greater displacement of the conus region than of the diaphragmatic part

The muscles of the right ventricle consist chiefly of trabeculae, the rest of the wall being fairly thin (Fig 92) When there is increased diastolic filling of the ventricle, it is mainly the crista supraventricularis and its two bands, as well as the

developed trabecular system than to thickening of the actual wall muscles (Fig 94). The trabeculation of the left ventricle is also powerful According to Edwards (219) the adult disproportion between the thickness of the two ventricles is usually established by both anatomic and electrocardiographic evidence by the end of the third ...

... to occur in the form of a squeezing movement, which starts at the apex and spreads toward the conus.

At the end of diastole, the conus has the appearance of a funnel, with its tip directed toward the *semilunar valves* (Fig. 437, B, p 470) It is wide in the frontal plane in particular and, with the possible exception of the semilunar valve region, its width always *exceeds* that of the pulmonary artery. In the contracted state, on the

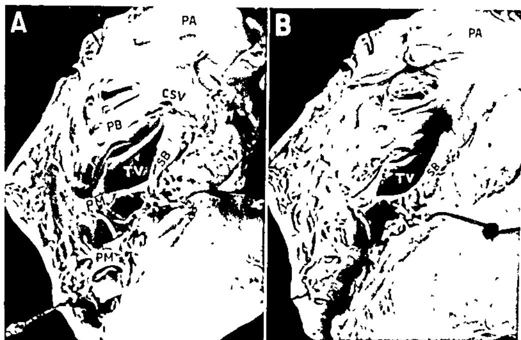


Fig. 92.—Right ventricle, normal case. Specimen has been mounted so that its details appear i[illegible]
c[illegible]
t[illegible]
t[illegible]
PB, parietal band, *PM*, papillary muscles, *SB*, septal band; *TV*, tricuspid valve



the cut surface D, severe pulmonary stenosis and endocardial vegetations, raised pressure in the right ventricle but no shunt, enormous hypertrophy of the wall muscles CSV, crista supraventricularis, I, infundibulum, PA, pulmonary artery, PB, parietal band, PM, papillary muscles, SB, septal band, TV, tricuspid valve

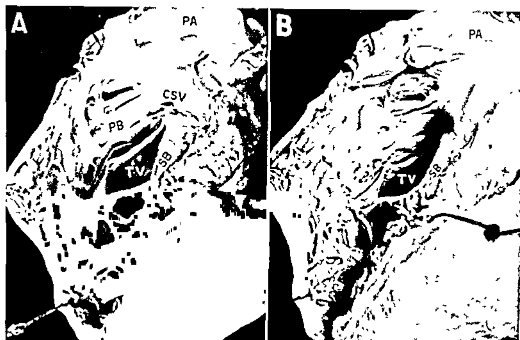


Fig. 92 —Right ventricle, normal case. Specimen has been mounted so that its details appea in essentially the same arrangement as on the lateral angiocardiogram of the right ventricle Th crista supraventricularis bulges slightly into the lumen, as do the septal and parietal bands Posi tion and direction of the large papillary muscles are seen in *A*, which is slightly rotated in rela tion to *B*, in order to show these details *CSV*, crista supraventricularis; *PA*, pulmonary artery *PB*, parietal band, *PM*, papillary muscles, *SB*, septal band, *TV*, tricuspid valve

contrary, it is tubular, and at the end of systole its width is less than that of the pulmonary artery (Fig. 437, *A*).

The *membranous part of the ventricular septum* lies farthest to the rear of the right corner, behind the medial cusp of the tricuspid valve and directly below the crista supraventricularis and its parietal band (Figs. 65 and 76). It is at this site that the majority of ventricular septal defects are found (*Fig. 93, C*)

In the proximal part of the *pulmonary artery* is a bulb-shaped dilatation, the *bulb* quently, in the lateral projection, the right branch is generally projected more caudally than the left and slightly in front of it (Fig. 544, p. 600). The right main branch over-rides the left atrium (Fig. 514).

As shown in Figure 95, the pericardial reflection lies high up on the main trunk of the pulmonary artery, slightly before the bifurcation When the pulmonary artery becomes dilated, the pericardium at its reflection does not become distended to a corresponding degree, but causes an indentation in the lumen (Fig. 185, p. 193).



Fig 95 —The pericardial reflection, which lies high up on the main trunk of the pulmonary artery, slightly anterior to the bifurcation *AO*, aorta, *PA*, pulmonary artery, *Per*, pericardium

of the pulmonary artery (Fig 509, p 565, Fig 425, p. 460) The *semilunar valves* are never opened so completely that they come into contact with the vessel wall. The blood between the valves and the wall prevents their adherence to it In normal cases, it is difficult to visualize the whole of the semilunar valves at angiocardiography, particularly during systole The valvular plane is usually easy to identify, owing to the "waist" which appears in the outline between the bulb and the infundibulum.

The right main branch of the pulmonary artery runs obliquely downward to the right, whereas the left main branch runs backward and somewhat to the left. Conse-

During ventricular systole, the valvular plane, together with the main trunk of the pulmonary artery, moves caudally, and there is concurrent dilatation of the artery, owing to the increased blood volume (Fig. 91) During diastole, the valvular plane returns successively to its original position.

Two of the *cusps of the pulmonary valve* border on the aorta, whereas the third lies anteriorly to the left. The designation of the cusps varies in the literature. In view of the embryologic development and of the often abnormal position of the great vessels in congenital heart disease, we have considered it most suitable to denote them as the *right posterior, left posterior* (both of these







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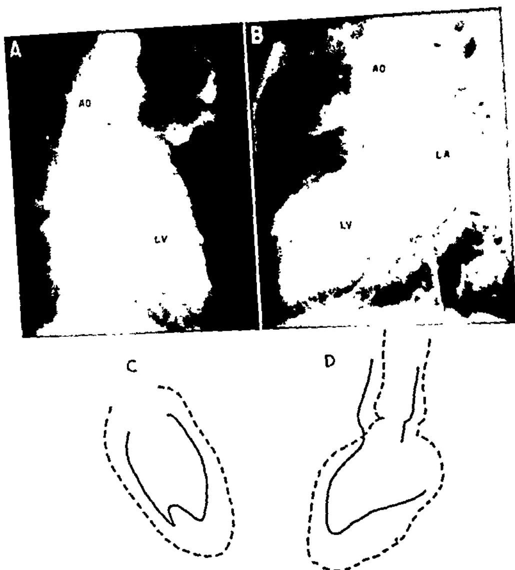


Fig 97 —Left ventricle, normal case. *A* and *B*, in diastole, its shape more rounded than that of the right ventricle, *C* and *D*, mode of contraction diastole (broken line), systole (solid line). The ventricle contracts fairly concentrically *AO*, ascending aorta, *LA*, left atrium, *LV*, left ventricle

bordering on the aorta) and the *anterior* cusp (Fig. 96).

As may be inferred from Figures 69, L–Q, and 97, the *left ventricle* has a more rounded form than the right. The band formation which is so typical of the right ventricle is lacking (Fig. 98). The trabeculation is mainly present in the apex region and along the left wall, i e., within the inflow tract. Both large *papillary muscles* (Fig. 98) arise from the anterior and posterior walls of the apex region. The septal surface, on the contrary, is largely smooth, and only a few thin muscular bands run

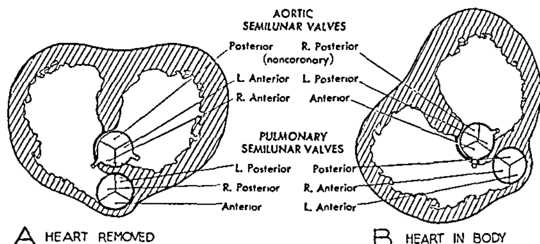


Fig. 96.—Diagrams showing bases for the two common systems of naming the semilunar [illegible] septum [illegible] (I N A [illegible] *Thomas,* Publisher, 1953].)

along it from the conus region toward the apex. The ventricular walls are thick and consist of a muscular mass with a largely circular course. This is presumably the reason why the left ventricle contracts more concentrically than the right (Fig 97) The contraction starts at the apex and spreads toward the conus (Fig. 99) During systole, a retraction of the wall in the posterior left part of the apex region is seen in many cases It is probably due to contraction of the coarse trabeculae which run from the region of the mitral ring toward the apex.

When the left ventricle is obliged to deal with a greater blood volume than normally, e.g., in patent ductus arteriosus, it seems to be mainly the trabeculae that become hypertrophied (Fig. 100). If, on the other hand, it has to work under raised pressure, e.g., in aortic stenosis, hypertrophy is more or less confined to the actual musculature of the wall (Fig. 100).

The *conus* is short and has no distinct borderline in the caudal direction (Fig 69). It is delimited medially by the septum, laterally and posteriorly by the anterior cusp of the mitral valve and anteriorly by the anterior wall of the ventricle. The membranous part of the septum lies inferior to the commissure between the right anterior and the posterior aortic cusps (Figs. 46 and 81). During systole, the conus and the semilunar valves move caudally to a lesser degree than on the right side. The movements of the apex region toward the conus are, on the contrary, more marked than on the right side. In the beginning of diastole, when filling of the ventricle is rapid, the anterior cusp of the mitral valve is displaced forward and to the right This causes *narrowing of the infundibulum, as can be* inferred from Figures 69, 99, and 101. As the blood flow to the ventricle ebbs, a gradual dilatation of the infundibulum takes place, and becomes maximal in the first part of systole (Fig. 101)



Fig 99 —Mode of contraction of left ventricle in the dog, conditions are similar to those in man At the S wave, a change in shape is seen, particularly in the apex region (*C*) *AO*, aorta, *LA*, left atrium *LV*, left ventricle

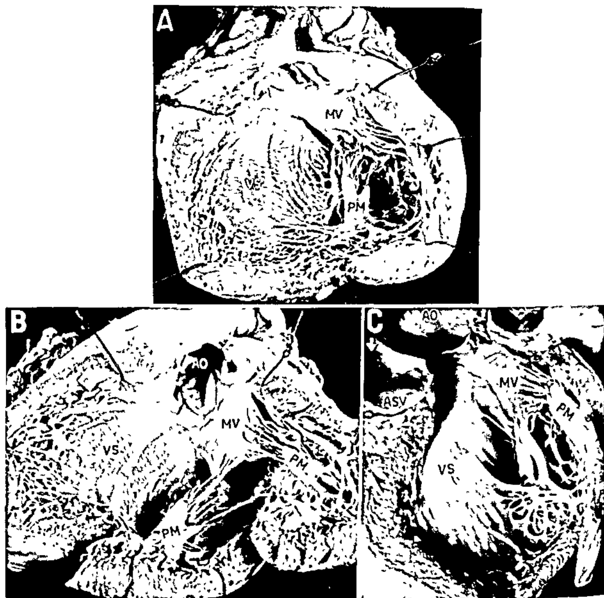


Fig. 98.—Left ventricle *A* and *B*, normal case. The septal wall is almost smooth. Only thi strands of trabeculae run from the cusps of the aortic valve toward the apex. Trabeculation i … sinus of Va … through th … ir valve, *M*\ mitral valve, *PM*, papillary muscles, *VS*, ventricular septum

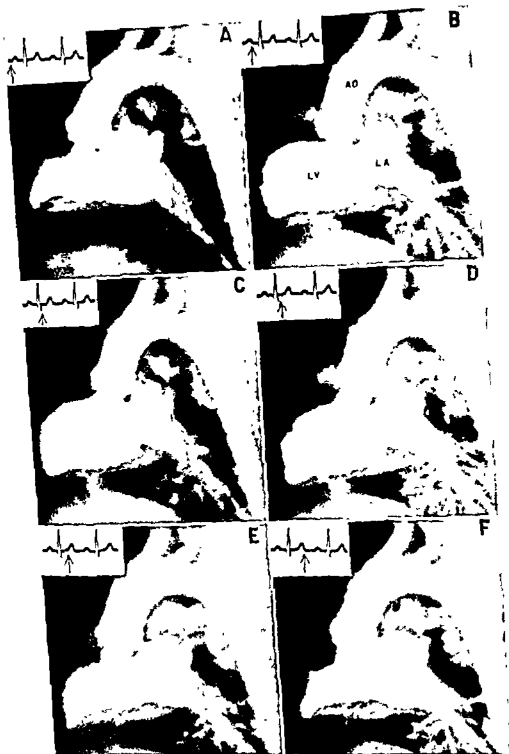


Fig 99 —Mode of contraction of left ventricle in the dog, conditions are similar to those in man At the S wave, a change in shape is seen, particularly in the apex region (C). AO, aorta, LA, left atrium, LV, left ventricle

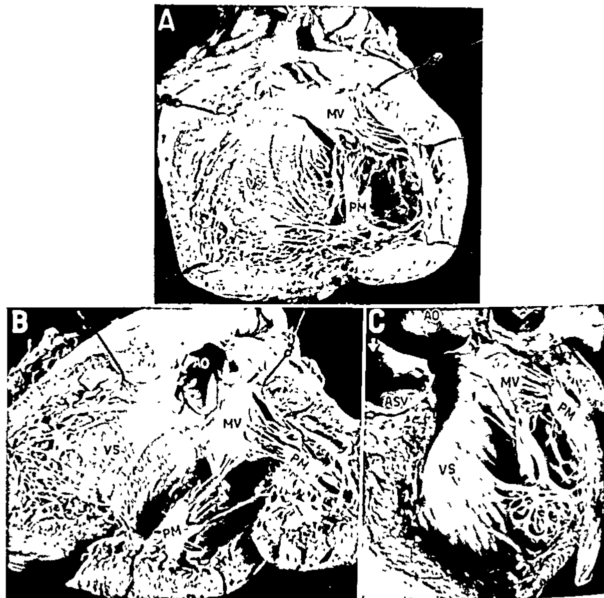
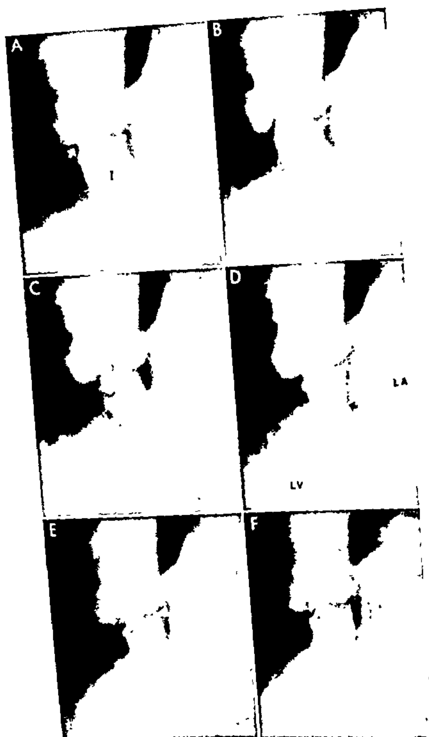


Fig. 98.—Left ventricle *A* and *B*, normal case The septal wall is almost smooth Only thin [illegible]

left ventricle has not yet left any marked traces *AO*, aorta, *ASV*, aortic semilunar valve; *MV*, mitral valve, *PM*, papillary muscles, *VS*, ventricular septum


A
B
I
C
D
LA
LV
E
F

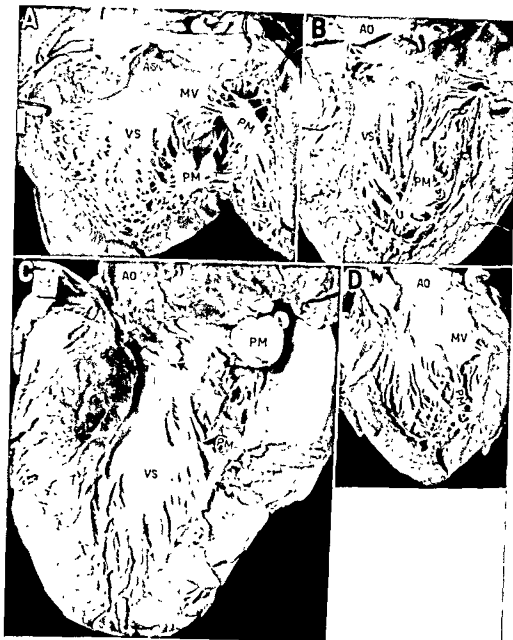


Fig 100.—Appearance of left ventricle *A*, atrial septal defect, normal trabeculation and thickness of the wall *B*, ventricular septal defect (arrow) with left to right shunt. distinct hypertrophy of trabeculae but no evident hypertrophy of the wall muscles *C*, [illegible] enormous hypertrophy of the wall muscles hypertrophy of the trabeculae is marked *D*, [illegible] ductus arteriosus with [illegible] pulmonary [illegible] appearance is the same as in *B* *AO*, [illegible] *ASV*, aortic semilunar valve, *I*, infundibulum, *MV*, mitral valve, *PM*, papillary muscles, *VS*, ventricular septum.

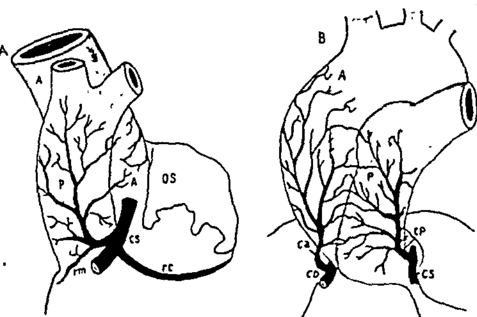


Fig 102 —The vasa vasorum of the aorta and pulmonary artery. *Left*, cardiopulmonary ar-
y, *right*, cardio-aortal and cardiopulmonary artery and the numerous anastomoses between
se two vessels *A*, aorta, *ca*, cardio-aortal artery, *CD*, right coronary artery; *CP*, cardiopulmo-
ry artery, *CS*, left coronary artery, *OS*, left atrium, *P*, pulmonary artery, *rc*, circumflex branch,
, myocardial branch (Redrawn from Muratori, G, Arch. sc med 82.24, 1946)

The proximal part of the *aorta*—like that of the pulmonary artery—presents a bulb-shaped dilatation, the *bulb of the aorta* (Fig. 574, p. 629). Under normal conditions, the aortic root lies dorsally to the conus region of the right ventricle. Consequently, in the frontal view at angiocardiography, it is entirely or partially overlapped by this region. At the end of ventricular systole, or when the right segment of the crista supraventricularis is displaced forward, the right segment of the aortic root is visualized in the angle formed by the infundibulum and the sinus region of the right ventricle (Fig. 269, p. 286). Two of the *cusps of the aortic valve* border on the pulmonary artery, and one lies posteriorly to the right (Fig. 96). For the same reasons as those brought forward earlier, we denote them as the *right anterior, left anterior* (both bordering on the pulmonary artery), and the *posterior* or *noncoronary* cusp.

THE CORONARY ARTERIES.—The *left coronary artery* arises in the wall of the left anterior sinus of Valsalva and runs laterally and ventrally between the root of the pulmonary trunk and the left atrium (43, 548). After a short distance, it divides into two branches: the *anterior descending branch*, which runs caudally in the anterior longitudinal sulcus as far as the apex of the heart, and the *circumflex branch*, which swerves round the base of the left atrium and runs in the coronary sulcus as far as the diaphragmatic surface of the heart (Fig. 3a). From these main branches are given off a number of small branches to the myocardium, the ventricular septum, the left atrium, and the aortic root. Anastomoses are also present, particularly between the circumflex branch and the right coronary artery (Fig. 3a, *B*).

The *right coronary artery* takes its origin from the right anterior sinus of Valsalva and runs laterally in the groove between the pulmonary conus and the right atrium (Fig. 3a). It then follows the base of the right atrium along the coronary sulcus and reaches the posterior longitudinal sulcus. Here, it divides into two branches: a large one, the *posterior descending branch*, which follows the posterior longitudinal sulcus, and a smaller one, which anastomoses with the circumflex branch of the left coronary artery. Several large branches are given off from the right coronary artery to the myocardium of the right ventricle, the ventricular septum, the right atrium, and the aortic root.

Like all the arteries of the human body, the coronary arteries may exhibit more or less marked anatomic deviations from the normal; they have been studied in detail by Guglielmo and Guttadauro (193, 194, 195), among others.

In hypertrophy of the ventricular musculature on one or the other side, dilatation of the corresponding coronary artery is generally observed, as a result of the increased blood flow. Conversely, in underdevelopment of one ventricle, a decrease in width of the corresponding coronary artery is seen.

The coronary arteries communicate with the rest of the systemic circulation through a system of anastomoses (195), via the nutrient arteries of the aorta and pulmonary artery, i.e., the cardio-aortal and cardio-pulmonary arteries (499) (see Fig 102)

THE CORONARY VEINS.—The coronary veins run parallel to the branches of the coronary arteries. They return blood to the right atrium via the coronary sinus.

The *great cardiac vein* originates in the epicardium of the anterior longitudinal sulcus (Fig. 3a). When it reaches the coronary sulcus, it swerves dorsally and follows the base of the left atrium. It empties into the distal end of the conorary sinus

The *middle cardiac vein* has its course in the posterior longitudinal sulcus It receives blood chiefly from the ventricular septum and the adjacent parts of the ventricular walls It empties into the coronary sinus slightly before the opening of the latter into the right atrium

The *small cardiac vein* lies in the coronary sulcus, at the base of the right atrium This vein also empties into the coronary sinus, close to its opening in the right atrium.

3 | Technique

CALIBRATED PHONOCARDIOGRAPHY

PHONOCARDIOGRAPHY involves the recording of the vibrations of the chest wall caused by the sound phenomena of the heart The wider clinical use of the method has been delayed by the technical difficulties, which are greater than those associated with electrocardiography. A prerequisite is for the method to be reproducible, in order to permit comparisons between tracings from the individual patient on different occasions and between the tracings from different patients The errors of the method are still far from negligible and are due to many factors, such as the variation in thickness of the thoracic wall and the difficulty in placing the microphone over the exact area of the maximal vibrations An international standardization of phonocardiography has been discussed on several occasions, but has not yet been effected

A more detailed technical description of earlier methods, as well as a presentation of the fundamental acoustic principles, has been given in previous papers They also include references to earlier works in this field (461, 463)

The apparatus used for recording the phonocardiograms in this book is constructed according to the principles reported in 1940 by Mannheimer (461). The filters are not, however, of the sharp cut-off type band-pass filters described there The method has been simplified by excluding the low-pass filters, so that our apparatus now contains only high-pass filters. This is because the decrease in amplitude of the heart sound phenomena with increasing frequency makes low-pass filters superfluous. Five different filters are used, we have designated them according to the frequency which actually appears with the highest amplitude in the records. These "standard frequencies" are 25, 50, 100, 200 and 400 cycles per second (cps). Thus, each standard frequency is situated 1 octave from its neighbor The frequency ranges actually recorded with the respective bands are: 12–50, 25–100, 50–200, 100–400 and 200–800 cps

In addition to these five filters, the apparatus is equipped with a filter having the same frequency response as the human ear. With this filter, a record is obtained which reproduces the different frequencies as they are heard on auscultation of the chest wall, i e , the "ear-like" tracing. The attenuation curves of the filters (1–6) are shown in Figure 104. Figure 105 indicates, on a linear scale, how different frequencies actually appear in the five bands, if the decrease in amplitude of the vibrations with rising frequency is taken into account. The curves in Figures 104 and 105 represent the frequency response of the whole apparatus from microphone to record

The microphone is of the ...

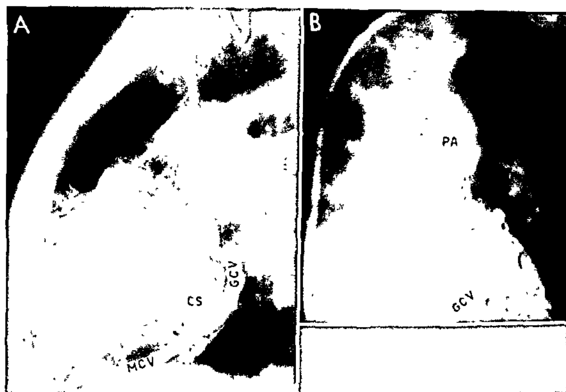


Fig. 103.—Venous anastomoses between coronary and systemic veins. Boy, aged 9 (K K 451030). A richly branched venous network spreads around the large vessels, it forms a communication between the coronary veins and the large venous trunks *CS*, coronary sinus, *GCV* and *MCV*, great and middle cardiac veins, *PA*, pulmonary artery.

There are also a number of small veins, such as the posterior vein of the left ventricle and the anterior cardiac veins, which empty either into the coronary sinus or its branches or else directly into the right atrium.

According to Ovenfors (521), a network of small veins spreads from the heart veins along the large venous trunks, they are in direct communication with the large venous trunks in the upper part of the mediastinum (see Fig. 103)

3 | Technique

CALIBRATED PHONOCARDIOGRAPHY

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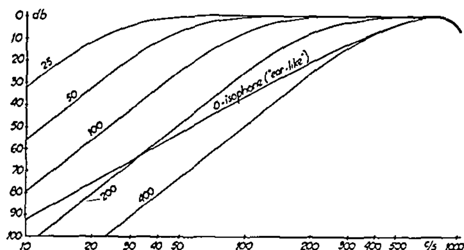
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The microphone is of the dynamic type,

which is more sensitive than the crystal microphone used earlier.

The phonocardiographic tracings reproduced in this volume were recorded with apparatus manufactured by the Elema Co, *Stockholm*. Two types were used. The larger tained with these two types of apparatus

A built-in device insures that the sensitivity of the apparatus remains constant. *The figure for the degree of amplification* gives the amplification in relation to the maximal amplification. In Figure 386, *A*



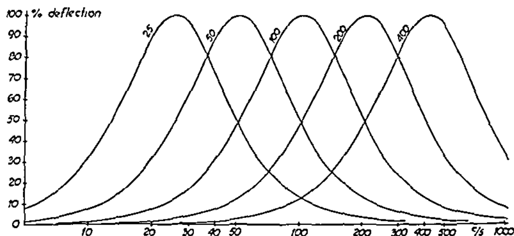


Fig. 105.—Calibrated phonocardiography. *The standard frequencies of the five frequency* bands

of them permits simultaneous recording of three channels and is connected to a six-channel electrocardiograph for photographic recording. The smaller apparatus has one phonocardiographic channel and is combined with an Elmqvist direct-writing jet-recorder electrocardiograph with two galvanometers. Identical tracings are ob- (p. 420), for example, the figure given is *1/20 and in Figure 386, C, it is 1/100* Equally high amplitudes in both figures thus designate sound phenomena that are five times as weak in the former case as in the latter This method is known as calibrated phonocardiography

RECORDING TECHNIQUE —All the chil-

dren were examined in the supine position and, if possible, when holding the breath. No sedatives were given.

In the case of infants and small children, a special chest piece was attached to the microphone. The microphone was supported by hand and light pressure exerted on the skin. A prerequisite for reproducible tracings is that the microphone be kept air-tight against the skin. On the other hand, excessive pressure will diminish the amplitudes (461). We have obtained better results with manual support of the microphone than with the application of rubber straps.

During the recording of pathologic sound phenomena, the microphone was consistently placed over the maximal point of audibility.

During the past few years, a more comprehensive phonocardiograph examination has been performed routinely. Recordings have been made over the following areas:

the apex,
the second left interspace (2nd L.I.S.)
the second right interspace (2nd R.I.S.),

and, if required, the third or fourth left interspace, or other areas over which significant auscultatory findings have been reported.

Great patience is required in the examination of small children. In some cases, satisfactory tracings can be made only when the patient is asleep.

The Phonocardiogram of Healthy Children

In most phonocardiograms of healthy children, four heart sounds and a faint systolic murmur can be recorded.

The *first heart sound* is caused by several factors, e.g., the closure of the atrioventricular valves (461). Its average duration is 0.10 sec, and it is rarely split. The early systolic click found in cases both of pulmonary dilatation and of aortic disease is not caused by the first heart sound, but originates from vibrations in the pulmonary artery and the aorta (417).

The *second heart sound* is of well established origin. It occurs on closure of the semilunar valves and marks the end of systole.

From the practical viewpoint, splitting of the second sound, i.e., its differentiation into one aortic and one pulmonary component, is of paramount importance. Leatham (416) showed, by means of simultaneous phonocardiographic recordings in the mitral and pulmonary areas, that in normal persons—most conspicuously in children—the second sound is always split over the pulmonary orifice. He found the first component to be due to closure of the aortic valve, and the second, to closure of the pulmonary valve. The splitting was most prominent at the end of inspiration. In left bundle-branch block, the aortic component occurs after the pulmonary one, but in nearly all other conditions—in both normal and pathologic cases—closure of the pulmonary valve invariably ends systole. If the pulmonary component is small and delayed, valvular pulmonary stenosis can be suspected. If its amplitude is increased, pulmonary hypertensio[illegible] ent. [illegible] mark[illegible] ally seen on the ECG.

The *third heart sound* is low pitched and therefore *is not, as a rule*, heard on auscultation. It can be recorded in about 60 per cent of normal children. It occurs in early diastole, about 0.13 sec after the beginning of the second sound and is due to muscular vibrations of the ventricular walls during the phase of rapid filling. It coincides with the descending limb of the *v* wave on the phlebogram. The third sound is best auscultated and recorded over the apex, with the patient in the left recumbent position.

The *auricular sound* is recorded about 0.12 sec after the beginning of the P wave on the ECG and is caused by atrial contraction. Under normal conditions it is recorded only below 100 cps, but in pathologic cases it may be higher pitched and of larger [illegible]

plitude. It is not heard on auscultation in normal cases.

The *physiologic systolic murmur* is, according to Levine (426), always of grade 1 or 2, i.e., "very slight" or "slight." It is best heard or recorded over the pulmonary orifice or lower down at the left sternal border, but sometimes it is distinctly audible over the apex or the whole precordium.

It is remarkable that, on the grounds of technical difficulties, most phonocardiographic studies give little information regarding the physiologic systolic murmur (see Fig. 388, A, p. 422). It is due to the high velocity of flow through the pulmonary valve. The stroke volume is approximately the same during exercise and at rest (345), whereas the ejection time of the ventricles is shortened during exercise, on heavy work it is only half that at rest (see Fig 117, p 121). This implies that the velocity of flow is doubled, thus producing a murmur of the same nature as that in atrial septal defect with a pulmonary flow twice the size of the systemic flow. In this form of heart disease, the ejection time of the right ven-

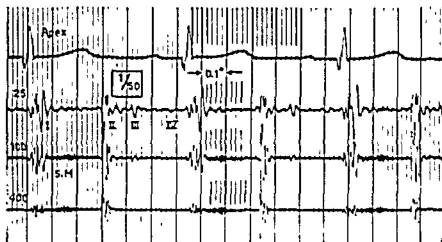


Fig. 106.—Phonocardiogram in a healthy child Boy, aged 5 (S E 471219) *I*, 1st sound, *II*, 2nd sound, *III*, 3rd sound, *IV*, auricular sound, *SM*, systolic murmur Boxed figure denotes degree of amplification, other figures denote standard frequencies of the filters

(454, 461). Improvements in the technique have now made it possible to obtain a true record of a faint systolic murmur in all children examined. The murmur is low pitched and has small amplitudes and does not, as a rule, reach the highest frequency range.

Mannheimer and Paulin (530) made the following observations in a series of 24 normal children. At rest and after the administration of dihydroergotamine, the shape of the murmur was that just described After exercise or the inhalation of amyl nitrite, the cardiac output increased, and the systolic murmur became stronger and protosystolic in time Its nature was similar to that found in atrial septal defect and in anomalous drainage of pulmonary veins tricle is normal, but, with a stroke volume that is twice as large as normal, the velocity of flow will be doubled

The normal phonocardiogram is illustrated in Figures 106 and 387 (p 421).

ELECTROCARDIOGRAPHY

The ECG was recorded with a six-channel photographic electrocardiograph, or sometimes with a direct-writing ECG apparatus with jet recorders (Mingograf) Both apparatus are manufactured by the Elema Co, Stockholm, according to Elmqvist's design

The standard leads, unipolar extremity leads and chest leads from V_{4R} to V_7 were recorded routinely Additional chest leads were sometimes used.

PHYSICAL WORKING CAPACITY

In evaluating the physical working capacity of patients with heart disease, it is highly important not to rely on statements in the history but to make objective tolerance tests. It is not uncommon for cardiac symptoms to appear after "heart disease" has been diagnosed by a physician, despite the presence of only a physiologic murmur. In children, it is especially difficult to obtain a correct idea of the degree of severity of the symptoms from the parents' statements alone As a rule the symptoms are exaggerated, but sometimes the reverse applies.

Various kinds of exercise tolerance tests have been used (112, 113, 509). We have used that of Sjostrand and Wahlund (607, 679) Several investigations have shown the merits of this method in the evaluation of both acquired (346, 679) and congenital heart disease (366), as well as in postinfectious myocarditis (55).

With this method, the physical working capacity is defined as the work, in kgm per minute, that can be performed by the patient sitting on a bicycle ergometer, with a pulse rate of 170 per minute, and in a relatively steady state The load is increased stepwise every six minutes, and the pulse rate is counted at rest and after two, four and six minutes' work at each load A steady state is considered to exist when the pulse rate does not increase more than 10 beats per minute during the last four minutes of each working period The load should be increased until the pulse rate reaches 150 to 170 per minute The initial load should be chosen so that the patient can carry out work at two or three different loads For healthy adult men in ordinary training, it is usual to choose 300, 600, and 900 kgm per minute, and for women, 200, 400, and 600 kgm per minute Since a linear relation exists between pulse rate and work load, the patient's physical working capacity can be obtained by interpolation or extrapolation (see Fig 107). If a steady state is not reached, the working capacity is calculated as a somewhat lower value than the greatest load carried out by the patient, provided that the pulse rate has reached 160 to 170 beats per minute.

The respiration rate is determined at rest and after three minutes' work at each load For recording of the electrocardiogram, the chest leads are used, with the reference electrode placed on the forehead (609). Recordings are made after five minutes' work at each load, as well as before the test, immediately after it is ended, and four minutes later. An orthostatic test is also made,

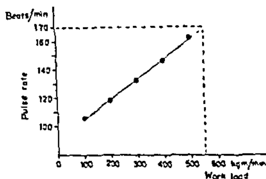


Fig. 107.—Exercise tolerance test in a healthy boy, aged 10 The rise in pulse rate is in linear relation to the increase in work load The physical capacity, defined as the work in kgm/min performed at a pulse rate of 170/min, is obtained by extrapolation. In this case, the working capacity was 550 kgm/min

the ECG and pulse rate being recorded after eight minutes in the erect position.

The exercise tolerance test is discontinued if subjective discomforts are felt or if electrocardiographic changes appear. In aortic stenosis, the test is not continued to a higher pulse rate than 130 to 150 per minute In cases with a large left to right shunt, it is often impossible to continue the test beyond a pulse rate of 150 per minute, since the fall in arterial oxygen saturation is so gre[illegible]

[illegible] the maximum tolerance is not reached at a pulse rate of 170 per minute. At this pulse rate, the oxygen intake in a young healthy man in good training is about

plitude. It is not heard on auscultation in normal cases.

The *physiologic systolic murmur* is, according to Levine (426), always of grade 1 or 2, i.e., "very slight" or "slight." It is best heard or recorded over the pulmonary orifice or lower down at the left sternal border, but sometimes it is distinctly audible over the apex or the whole precordium.

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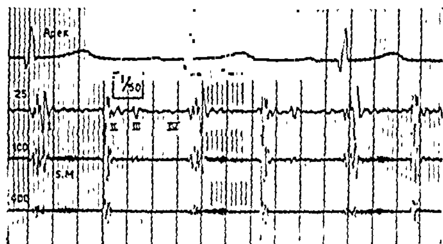


Fig. 106.—Phonocardiogram in a healthy child. Boy, aged 5 (S.E. 471219) *I*, 1st sound; *II*, 2nd sound, *III*, 3rd sound, *IV*, auricular sound, *SM*, systolic murmur. Boxed figure denotes degree of amplification, other figures denote standard frequencies of the filters.

The normal phonocardiogram is illustrated in Figures 106 and 387 (p. 421).

ELECTROCARDIOGRAPHY

The ECG was recorded with a six-channel photographic electrocardiograph, or sometimes with a direct-writing ECG apparatus with jet recorders (Mingograf). Both apparatus are manufactured by the Elema Co., Stockholm, according to Elmqvist's design

The standard leads, unipolar extremity leads and chest leads from V_{4R} to V_7 were recorded routinely Additional chest leads were sometimes used

sels and one harder for a study of such details as the retrocardial area, the left atrium, the aorta, and abnormal vessels. In addition, two exposures were made in oblique projections (one in each direction), as well as one in the lateral projection. Barium sulfate was given by mouth to all the patients, including those examined in the recumbent position, to permit visualization of the esophagus

CRITERIA

The roentgenologic diagnosis of congenital heart disease is based to a great extent on the evaluation of the size and position of the individual chambers of the heart. This may, however, be subject to considerable difficulties, since their borderlines may be hard to identify Enlargement of one chamber may be compensated for by underdevelopment of another and may therefore escape attention. Enlargement is nevertheless usually associated with a change in the configuration, which can be used as indirect evidence. A decrease in volume of one of the chambers is almost invariably difficult to establish directly. Anomalies of position may also hamper the assessment

Fluoroscopic examination plays an important role in the investigation of congenital heart disease It aids in the evaluation of the size and relative position of the individual cardiac chambers Moreover, certain conclusions—even if they are limited —can be drawn regarding the hemodynamics by means of observation of the pulsations of the heart and great vessels In our opinion, a study of the pulsations of the atria and of the great vessels is of particular diagnostic value

The normal *right atrium* occupies the right segment of the heart and, in the frontal view, forms its right border Its superior portion does not, as a rule, extend to the level of the carina Normally, there is no sharp borderline between the right auricular appendage and the rest of the atrium In atrial enlargement, the lateral and anterior surfaces exhibit increased curvature, so that the atrial border is displaced laterally and the superior part of the appendage is shifted cephalad, not infrequently far above the carina (Fig 633, p. 701). A distinct borderline is often seen between the appendage and the rest of the [illegible]

the fact that another chamber is underdeveloped and the atrium partly occupies its site, enlargement can still be suspected on the grounds of the change in shape. We have the impression that enlargement must be considerable before it can be definitely established

Because of its purely frontal position, the *right ventricle* is best judged in the lateral projection. Normally, only part of its surface is contiguous to the anterior thoracic wall In the presence of dilatation, this contiguity is increased, and may be considerable, partly as an effect of dilatation of the infundibulum (Fig. 392, p 425). The dilatation may, however, be overestimated in this projection if an enlarged right auricular appendage is added to the cephalic segment of the ventricle (Fig. 146, p. 159). It must also be borne in mind that in children with a flat thorax, the heart lies more vertically, and this results in a greater contiguity to the anterior thoracic wall. In the youngest children, the evaluation is not infrequently hampered by the large thymus, which fills the retrosternal space above the heart.

In the frontal projection as well, dilatation can be demonstrated if it is marked and includes the infundibulum, which then bulges in the superior part of the left border (Fig. 520, p 574). The border therefore has an angulated appearance.

In right ventricular *hypertrophy, there is increased curvature of the anterior surface of the heart. With* a considerable rise in pressure in the ventricle, it may be very conspicuous *(Fig 151, p. 164)*. When hypertrophy chiefly involves the sinus region, the diaphragmatic surface of the heart presents an increased curvature; this results in upturning of the apex (Fig. 235, p. 258). In this event, *the right ventricle* often forms

80 per cent of the maximal oxygen intake (29).

The adult patients were investigated at the Department of Clinical Physiology, where an electrically braked bicycle ergometer (347) was used. This ergometer has a *constant load at various pedaling rates.* The children underwent the tests at our laboratory, where a friction ergometer was used.

In healthy individuals, there is a good correlation between the physical working capacity, the total amount of hemoglobin of the body, and the heart volume in the recumbent position. The correlation between body surface area or body weight and working capacity and heart volume, respectively, is poorer (345, 398), especially in adults. In order to evaluate the physical working capacity more accurately, both the heart volume and the total amount of hemoglobin or blood volume (144, 608) should be determined, and the predicted value of the physical working capacity can then be obtained from the regression equation in healthy individuals of varying age and sex (345). These determinations were made in all adult patients for whom the working capacity is given, but not in the children In the latter, the physical working capacity is expressed in relation to the body weight and compared with the normal values given by Bengtsson (54).

During exercise in a steady state, when large groups of muscles are involved, the working capacity is limited by the oxygen-transporting ability of the circulatory system provided that respiration and neuromuscular function are normal This can be expressed as the oxygen intake per heart beat (oxygen pulse) at the maximal pulse rate which can be maintained in equilibrium. The working capacity is then determined by the stroke volume and the average utilization of the oxygen of the blood in the systemic circulation during exercise. The physical working capacity, determined by the method described, therefore shows good correlation with the stroke volume, both in healthy subjects and in those with heart disease (346).

ROENTGENOLOGIC EXAMINATION

Plain Film Roentgenography

All children who were unable to stand were examined in the recumbent position with the arms stretched above the head, which results in an artificial state of inspiration. On the grounds both of the rapid heart rate and of the fine caliber of the pulmonary vessels, a very short exposure time was used (0.003 sec), as well as a high current intensity (600 ma). The target-to-film distance was 1.25 meters (50 in.) and the voltage 55 to 75 kv (three-phase apparatus). Attempts were also made to examine these children in both the hanging and the sitting position The roentgenograms taken in the recumbent position were, however, generally more satisfactory.

A slightly different technique was used for those patients who could stand and were examined in the erect position with the arms stretched above the head For the youngest children, the exposure time was still 0.003 sec, but for older children it was increased to 0 01 sec and for adults to 0 028 sec. The current intensity was, on the contrary, decreased to 350 ma and the voltage raised to 150 to 200 kv. Target-to-film distance was 1.5 meters (62 in.) and the filter used 6 to 8 mm aluminum.

A secondary grid was not used, since it hampers the evaluation of the size and course of the small pulmonary vessels. Instead, use was made of an air layer of 10 cm between the patient and the film, of tinfoil 0.1 mm thick, or of a steel plate 0 8 mm thick The last usually gave the best results This was because the distance from the film to the patient was shorter, and harder radiation could be used.

In order to obtain the optimal detail, only finely-grained intensifying screens (Siemens Diamant) were used in all examinations of the heart and lungs.

The roentgen apparatus and tubes were of the same manufacture and type as stated on page 124

Two frontal pictures were taken routinely, one softer for evaluation of the ves-

sels and one harder for a study of such details as the retrocardial area, the left atrium, the aorta, and abnormal vessels. In addition, two exposures were made in oblique projections (one in each direction), as well as one in the lateral projection. *Barium sulfate was given by mouth* to all the patients, including those examined in the recumbent position, to permit visualization of the esophagus.

Criteria

The roentgenologic diagnosis of congenital heart disease is based to a great extent on the evaluation of the size and position of the individual chambers of the heart. This may, however, be subject to considerable difficulties, since their borderlines may *be hard to identify* *Enlargement of one* chamber may be compensated for by underdevelopment of another and may therefore escape attention Enlargement is nevertheless usually associated with a change in the configuration, which can be used as indirect evidence A decrease in volume of one *of the chambers is almost invariably diffi*cult to establish directly Anomalies of position may also hamper the assessment.

Fluoroscopic examination plays an important role in the investigation of congenital heart disease It aids in the evaluation of the size and relative position of the individual cardiac chambers Moreover, certain conclusions—even if they are limited —can be drawn regarding the hemodynamics by means of observation of the pulsa*tions of* the heart and great vessels. In our opinion, a study of the pulsations of the atria and of the great vessels is of particular diagnostic value

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In right ventricular hypertrophy, there is increased curvature of the anterior surface of the heart With a considerable rise in pressure in the ventricle, it may be very conspicuous (Fig 151, p. 164) *When hy*pertrophy chiefly involves the sinus region, the diaphragmatic surface of the heart presents an increased curvature, this results in upturning of the apex (Fig. 235, p. 258). In this event, the right ventricle often forms

the actual apex and gives it a fairly charac-

of the two ventricles, a notch in the border appears at the level of the ventricular septum (Fig. 235, p. 258). This is most distinct when the left ventricle is small.

The *left atrium* is, as a rule, the chamber of the heart that is easiest to evaluate. In view of the purely dorsal position of the atrium, it is visualized best in the frontal and lateral projections. Normally, except during the first days of life, it has a straight or only inappreciably bulging dorsal border. Estimation of its shape is facilitated by filling of the esophagus. It must, however, be recalled that in enlargement of the atrium, the esophagus is also displaced laterally. The part of the atrium lying most dorsally is then often seen behind the esophagus; consequently, the degree of dorsal displacement of the latter is not representative of the maximal expansion of the atrium. In enlargement of the right ventricle, the heart is sometimes tilted backward, so that the course of the esophagus is curved and simulates slight atrial enlargement.

The left auricular appendage extends forward and medially and lies beside the pulmonary artery, usually forming a short segment of the border between the main trunk of the pulmonary artery and the left ventricle. This is the only part of the normal atrium that can be identified in the frontal projection. In dilatation, the shape of the atrium is more rounded, it is often depicted as a distinct shadow projected within the heart, and its right border in particular is conspicuous. If atrial dilatation is marked, the right border of the heart is formed partly by the left atrium, and the angle of tracheal bifurcation is increased owing to lifting of the bronchi.

Of all the chambers of the heart, the *left ventricle* is the most difficult to judge. This is because its borderlines are to a great extent hard to identify. In oblique projections, we have never been able to identify the interventricular groove with certainty. According to Taussig (650), the size of the left ventricle is best evaluated in the left oblique projection, by determination of the position of the ventricle in relation to the spine. Great difficulties are, however, involved in standardizing the projection, owing to the influence of the body build on the position of the heart. Moreover, when there is right ventricular enlargement, the left ventricle is displaced dorsally and is then erroneously regarded as enlarged. In the frontal projection, the interventricular groove can not infrequently be visualized in certain types of heart disease associated with right ventricular hypertrophy, in the form of an indentation at the apex (Fig. 235, p. 258; Fig. 396, p. 428). The position of this indentation does not, however, allow any definite conclusions regarding the size of the left ventricle.

The shape of the ventricle provides certain information about its size. In hypertrophy, when the increase in volume is not as a rule marked, the left border of the heart presents increased curvature and the apex is broad (Fig. 543, p. 599). In dilatation, especially when it is severe, the left border forms—on the contrary—a long, gradual slope and the apex cannot be visualized without superimposition of the diaphragmatic dome (Fig. 599, p. 660).

The *heart volume* was calculated according to the formula of Rohrer and Kahlstorf and correlated to the body surface area. The normal values for the respective age groups were taken from Carlgren and Eek (143), Kjellberg *et al.* (394) and Lind (436).

ELECTROKYMOGRAPHY

The electrokymograph registers the changes of roentgen ray absorption of the chest due to heart action and breathing. The principle of the apparatus is the transfer by a fluorescent screen of the roentgen rays to light in a photomultiplier tube. The output current from the tube is filtered, amplified, and fed to a recorder. This modern type of electrokymograph was introduced by Henny and Boone (331) in 1945. For clinical purposes, it is used almost exclusively to record the pulsations of the

heart and great vessels. These are complex, and changes both in volume and in position are reflected on the electrokymogram.

We used two electrokymographs; one, manufactured by the Swedish Philips Co., was described in detail in 1949 (239); the other was made by AB Elema-Jarnh, Stockholm In order to counteract variations in the main voltage, balanced phototubes are time-constant influence on the electrokymogram

As a reference tracing, we consistently used the electrocardiogram, lead II, and sometimes the phonocardiogram as well; occasionally, the apex cardiogram was also used.

The phonocardiogram was obtained with a piezoelectro-microphone and a three-link

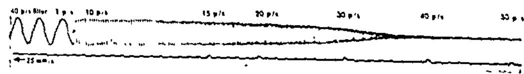


Fig 108.—Filter response curve of the electrokymograph amplifier.

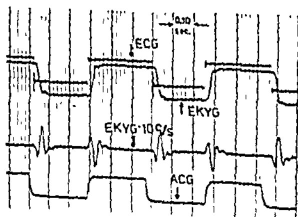


Fig 109.—
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used in the latter apparatus The frequency response of the amplifier of this electrokymograph is as satisfactory as theoretically possible, i e , the filter cuts frequencies above half the carrier frequency, which for full-wave roentgen ray apparatus is 100 cps, Figure 108 shows the actual frequency response of the electrokymograph amplifier Frequencies around 10, 15, and 20 cps, which are hidden in the electrokymogram, are filtered out by four-link C R high-pass filters, three in number (This matter will not be entered into here) It is evident from Figure 109 that the electrokymograph is d-c-connected which implies that there is no

C R filter, giving a frequency response corresponding to the human ear

$$(CR = \frac{1}{2\pi \cdot 400} \text{ sec}).$$

The microphone is partly translucent to roentgen rays, which facilitates its use.

The apex cardiogram was obtained with a receptor of condenser type that gave with the input grid leak a time constant of 1.5 sec

The electrokymograph filter introduces a time lag of 0.01 sec between the electrocardiogram and the electrokymogram (Fig. 109). No lag exists between the electrocar-

diogram, phonocardiogram, and apex cardiogram.

As a rule, the border electrokymograms were recorded; sometimes the density curves were also used. When the former are recorded, the slit of the photocell should be adjusted so that the border pulsates in its center. The segment to be studied should, as far as possible, be visualized without superimposition of other structures. Interference tracings recorded in the borderline areas between different chambers of the heart or the vessels are also difficult to interpret. Only central rays should be used for the recordings, a small diaphragm is therefore desirable. It is suitable to use a relatively high current intensity (if necessary, up to 5 to 6 ma) and low voltage, about 70 kv. For adjustment of the photocell, 2 ma usually suffices.

Routinely, the electrokymograms were recorded with the patient sitting. In simultaneous recording of the pressure curves and the electrokymograms, the patient was in the recumbent position. Children under 5 years are seldom suitable subjects for electrokymographic examination.

Surveys of electrokymographic findings under normal and pathologic conditions have been reported by, among others, Boone *et al.* (80), Luisada and Fleischner (444), Willis *et al.* (704), Dussaillant *et al.* (213), and Dack and Paley (173, 174) The method seems seldom to have been applied in the investigation of *congenital* heart disease.

In our series, the tracings of greatest diagnostic value were those from the pulmonary artery and the aorta, followed by those from the right atrium, whereas those from the ventricles and the left atrium were seldom helpful. It was endeavored to base their interpretation on the direction, slope, and duration of certain components of the curves and to refrain from referring to the amplitude other than in comparison with other segments of the electrokymogram. The time data in relation to the reference tracings are corrected for the time lag in the filter of the electrokymograph

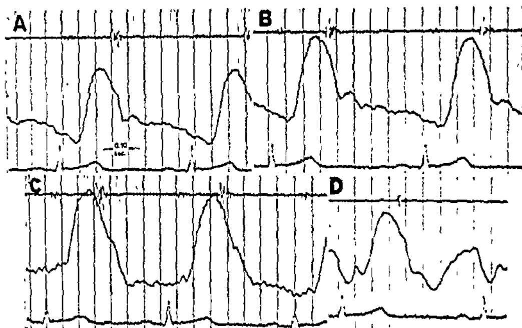
NORMAL ELECTROKYMOGRAMS

Descriptions of normal electrokymograms are found in the aforementioned works, among others. In the following account, only their general appearance is discussed, with particular reference to the relation to the phonocardiogram. A detailed study of this matter has been made by Reinhold and Rudhe (552). Current circulatory physiologic terms are used All the tracings shown, except those in Figure 111, *A–C*, are from the same individual, a 16-year-old boy (B.K. 380201).

The electrokymograms of the great vessels have essentially the same shape as volume curves and intra-arterial pressure curves.

In the *pulmonary artery electrokymogram* (Figs. 110 and 111), the average onset of the rise is 0.10 to 0.14 sec after the Q wave. The slope is steep, with an average duration of 0 12 sec, before the gradient changes and the curve definitely deviates toward the summit. This is reached after the middle of systole, but never at its end, and a distinct vertex is generally seen. At the end of the phase of reduced ejection, an inflection marks the transit into the protodiastolic phase. This has a duration of 0 03 to 0.05 sec and is ended by an incisura and dicrotic wave. These lie in a variable segment of the descending limb, but normally never occur in the immediate vicinity of the peak The incisura occurs 0 01 to 0.02 sec after the pulmonary component of the second sound. Postdicrotic waves are common Variations in the appearance of the curves are seen in recordings over different segments of the pulmonary artery(Fig. 110). Good agreement is found between the electrokymogram and the intra-arterial pressure curves registered simultaneously (Fig 111) The relation to the apex cardiogram may also be inferred from Figure 111 In tracings from the hilar vessels, the upstroke takes place later than over the main trunk, and the incisura and dicrotic wave are usually less distinct.

The *aortic electrokymogram* differs somewhat in appearance when it is re-



sec of the pulmonary component of the 2nd ... ejection and incisura. Incisura occurs within 0.01
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... 0.04–0.05 sec.

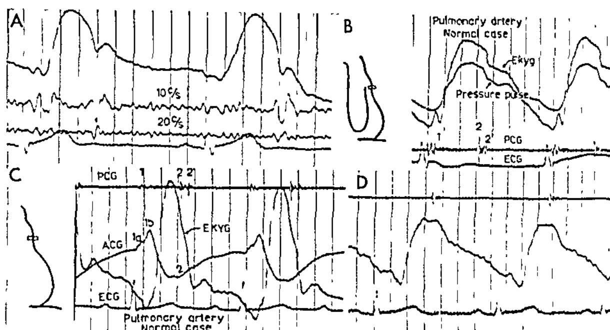


Fig. 111.—Normal pulmonary artery electrokymograms. *A*, lower part of "pulmonary arc." Middle curves are frequency channels around 10 and 20 cps filtered out of the electrokymogram and amplified. *B*, electrokymogram *(EKYG)* recorded simultaneously with pressure pulse, phonocardiogram *(PCG)* and electrocardiogram *(ECG)*. *PCG* over pulmonary area 2 and 2', aortic and

monary component of the 2nd sound. Time lag in pressure recording, 0.01 sec. *C*, electrokymogram *(EKYG)* recorded simultaneously with the apex cardiogram *(ACG)*, *PCG* and *ECG*. Onset of rise, 0 01 sec after *1a* in *ACG*, which marks closure of atrioventricular valves. *1b*, opening, and 2, closure of aortic valve Small extra wave at onset of rise. Duration of protodiastolic phase, 0 05 sec. Incisura occurs 0 02 sec after the pulmonary component of 2nd sound. Distinct dicrotic wave. *D*, electrokymogram of left hilum. *PCG* over pulmonary area Onset of rise 0 16 sec after Q wave. *Duration of rise, 0.13–0.14 sec. Incisura 0 05 sec after 2nd sound.*

corded over the ascending aorta and the aortic arch, respectively.

In the *ascending aorta* (Fig. 112) the onset of upstroke is 0.09 to 0.14 sec after the Q wave. The upstroke has a steep slope and ends in a peak or a distinct inflection. The average duration of this segment of the systolic limb is 0 11 sec. The incisura can almost consistently be demonstrated and recorded 0 01 to 0.02 sec after the second aortic sound. The "dicrotic" wave is composed of a smaller initial wave followed by a larger *one*

In the *aortic arch* tracing (*Fig 112*), the onset of rise is 0 01 *to* 0 02 sec later than in the electrokymogram of the ascending aorta, but the delay may *amount to* 0 04 sec. The upstroke seldom lasts for more than 0 11 sec and, after an inflection, passes in a "saddle shape" into the crest of the curve. The protodiastolic phase and Incisura are generally distinct and occur at a varying interval after the second aortic sound. The dicrotic wave is less conspicuous than in tracings from the ascending aorta. It overlaps the uppermost part of the descending limb.

Minor waves in presystole and after QRS occur in the electrokymogram of both the pulmonary artery and the aorta; they are *caused by atrial contraction and the isometric* contraction of the ventricles.

The *atrial electrokymogram* (Fig 113) is composed of three positive main waves, *one presystolic*, one *systolic* and one systodiastolic, and of a varying number of smaller, superimposed waves. *In the* presystolic part of the wave, *the* negative deflection repre-

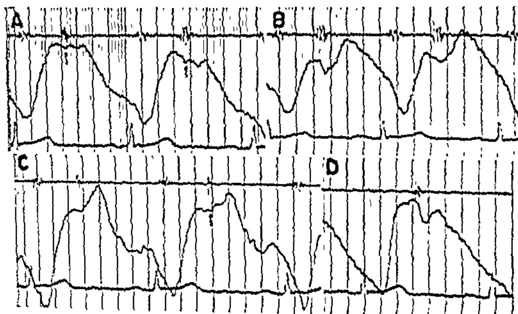


Fig 112 —Normal [illegible] wave.

sents atrial contraction. It starts 0.12 to 0.14 sec after the beginning of the P wave and has a duration of 0.04 to 0.08 sec. Under normal conditions, it seldom amounts to 0.09 sec and practically never to 0.10 sec. The course of the deflection nevertheless varies; it may be directed horizontally or upward and cannot invariably be exactly delimited. In the systolic wave, the ascending limb is related to the bulging of the valve during the ventricular phase of isometric contraction, and the descending limb to the movement of the atrioventricular plane during the phase of ejection The latter limb has a minimum duration of 0.05 sec. The systodiastolic wave is to be ascribed mainly to filling and emptying of the atria.

There are only small differences between the tracings from the two atria The most important of them is that ventricular influence during systole is more apparent on the right side.

Two features are essential for evaluation of an impediment to emptying of the atria One is the duration of atrial contraction, which then amounts to 0.10 sec or more. The other is the position of the lower points of inflection in the atrial phases of emptying. Normally, they are recorded at approximately the same level. In impaired emptying, the presystolic end-point is distinctly lower than the other.

The *ventricular electrokymograms* bear considerable resemblances to the volume curves recorded cardiometrically. They are, however, far from directly comparable, and the electrokymograms seldom allow reliable conclusions to be drawn with respect to emptying and filling conditions of the ventricles. The curves vary in appearance when they are recorded over different segments of the ventricles (239)

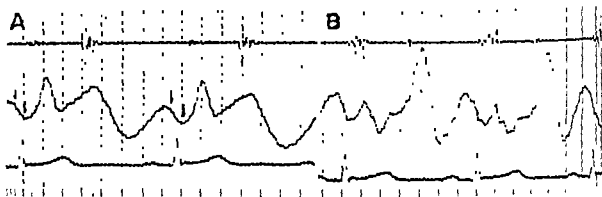


Fig. 113.—Normal atrial electrokymograms *A*, right atrium. Phonocardiogram over the pulmonary area. Atrial contraction (between arrows) starts 0.14 sec after beginning of P wave Duration of atrial contraction, 0.07 sec, the corresponding deflection passes the peak of the R wave. A semihorizontal segment follows, broken by an inflection at the main vibrations of the 1st sound, corresponding to closure of the A-V valve. Ascending limb of the following wave is synchronous with the phase of isometric contraction of the right ventricle, and its descending limb with the first part of the ejection phase of the ventricle. In the subsequent wave, expansion of the atrium ensues first, then emptying, which continues until the point marking onset of the next wave *B*, left atrium. PCG over apex Tracing of essentially the same appearance as in *A*

In principle, they are composed of a negative, systolic deflection and a positive deflection during diastole (Fig. 114). The onset of the ejection phase seldom coincides with the uppermost notch recorded after the QRS complex, but with an incisura below it. The protodiastolic phase and isometric relaxation can often, but far from invariably, be identified

In tracings from the infundibular region of the right ventricle, the systolic deflection is steep. The isometric phase of relaxation is marked by a peak, its size varying in recordings over different segments of the infundibulum.

CARDIAC CATHETERIZATION

Since Cournand and Ranges (166) in 1941 introduced cardiac catheterization as a clinical method, it has been used routinely in the diagnosis of congenital heart disease as well. Detailed descriptions of the technique have been given by other writers (45, 61, 163, 188, 202, 463, 620, 711, 712). Consequently, the following will be confined to a description of our apparatus and a discussion of certain aspects of particular importance in the catheterization of children.

APPARATUS FOR PRESSURE RECORDING

During the first years of our work, we used a condenser manometer according to Hansen (316) (manufactured by Kaiser's Laboratorium, Copenhagen). Thereafter we used a strain gauge manometer in combination with a carrier-frequency amplifier (manufactured by the Elema Co., Stockholm). The manometer is very stable and has a negligible zero drift. By means of a switch, it is possible to alter the sensitivity range without recalibration. The manometer is matched to the catheter by a capillary damping. The frequency response of the Elema manometer has been studied by Holmgren (344). He also tested our manometer, the results when using a no. 8 Lehman catheter are shown in Fig. 115.

The tracings were recorded with a six-channel photographic electrocardiograph of Elmqvist's design (manufactured by the Elema Co., Stockholm). The following recordings can be made: two pressure tracings, electrocardiogram, phonocardiogram, electrokymogram, apex cardiogram, venous

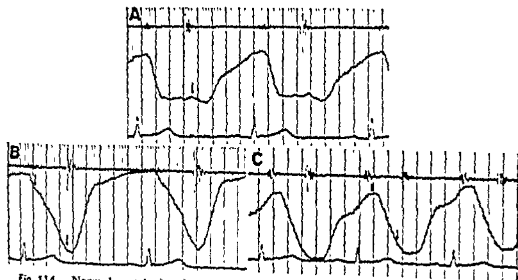


Fig. 114.—Normal ventricular electrokymograms. A, infundibulum of right ventricle. Phonocardiogram over the pulmonary area. Inward movement during later part of isometric phase of contraction and rapid ejection phase. Suggested outward movement ...

... phases.

sents atrial contraction. It starts 0.12 to 0.14 sec after the beginning of the P wave and has a duration of 0.04 to 0.08 sec. Under normal conditions, it seldom amounts to 0.09 sec and practically never to 0.10 sec. The course of the deflection nevertheless varies; it may be directed horizontally or upward and cannot invariably be exactly delimited. In the systolic wave, the ascending limb is related to the bulging of the valve during the ventricular phase of isometric contraction, and ing. Normally, they are recorded at approximately the same level. In impaired emptying, the presystolic end-point is distinctly lower than the other.

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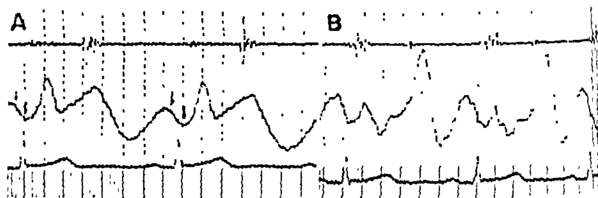


Fig. 113.—Normal atrial electrokymograms. *A*, right atrium Phonocardiogram over the pulmonary area. Atrial contraction (between arrows) starts 0 14 sec after beginning of P wave Duration of atrial contr... wave. A semihorizontal ... sound, corresponding to ... chronous with the phase of isometric contraction of the right ventricle, and its descending limb with the first part of ... the ventricle. In the subsequent wave, expansion of ... ich continues until the point marking onset of the next ... ng of essentially the same appearance as in *A*.

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In principle, they are composed of a negative, systolic deflection and a positive deflection during diastole (Fig. 114). The onset of the ejection phase seldom coincides with the uppermost notch recorded after the QRS complex, but with an incisura below it The protodiastolic phase and isometric relaxation can often, but far from invariably, be identified.

In tracings from the infundibular region of the right ventricle, the systolic deflection is steep The isometric phase of relaxation is marked by a peak, its size varying in recordings over different segments of the infundibulum.

venous blood, particularly in the presence of atrial septal defect or anomalous drainage of pulmonary veins. In patent ductus arteriosus, the blood in the pulmonary artery is not completely mixed. Moreover, considerable variations are often found in the oxygen saturation of blood from the same chamber of the heart from one moment to the next This applies in particular to infants and in certain severe cardiac malformations In transposition of the great vessels, the variations associated with respiration may be appreciable (see p. 787).

In cases with a large right to left shunt and a low blood flow through the pulmonary artery, numerous collaterals may have developed between systemic arteries and the peripheral branches of the pulmonary artery. The oxygen content of the pulmonary artery cannot then be used for calculation of the pulmonary blood flow (62)

In order to calculate the blood flows with some degree of accuracy in cases with shunts, it must be possible to take samples from systemic arteries, as well as distally and proximally to the shunt, simultaneously and during the greater part of the period in which the expiratory air is collected for calculation of the oxygen intake This requires introduction of an arterial catheter and of at least two heart catheters Such investigations were made in only a few older children and adults, and chiefly to study the effect of exercise They were performed at the Department of Clinical Physiology, the technique has been described earlier (366)

When the level of the shunt has been determined by the photometric blood-gas analysis method, the catheters are placed so that mixed venous blood can be obtained concurrently with samples from the pulmonary artery and systemic arteries Sampling of the blood is done slowly, during determination of the oxygen intake and recording of the pulse rate The expired air is collected in Douglas bags for 10 minutes at rest and four to five minutes during exercise, the oxygen and carbon dioxide being analyzed with the Haldane-Liljestrand apparatus. The gas volume is measured in a gasometer and the oxygen intake calculated.

After determination of the pressure and cardiac output at rest, an exercise test is performed in the same way as the usual exercise tolerance test, but in the recumbent position. The work load is increased stepwise and the cardiac output determined after four minutes' work at each load, when a relatively steady state has been reached. If the patient carries out three stepwise increased loads, the work lasts for a total 30 minutes. During this time, the pressure is measured continuously and simultaneously in the pulmonary artery, right ventricle, and brachial artery and sometimes in the right atrium as well.

The blood flows are calculated by the Fick method If the arterial oxygen saturation is normal, the pulmonary flow (Qp) is calculated as follows.

$$Qp\ (l/min) = \frac{\dot{V}_{O_2}}{C_{O_2}\ (\text{brach art.}) - C_{O_2}\ (\text{pulm art.})} \qquad (1)$$

$\dot{V}_{O_2}$ is the oxygen consumption in ml per minute, and C_{O_2} the oxygen content in ml per liter of the blood from the cavity given in parentheses.

If the arterial oxygen saturation is decreased, the oxygen content of the pulmonary venous blood is used in equation (1) instead of the systemic arterial blood If pulmonary venous blood cannot be obtained, the oxygen content is calculated from a predicted saturation of 97 per cent If it can be assumed that the oxygen saturation of the pulmonary venous blood is decreased, the oxygen tension of the arterial blood should be calculated during breathing of air and of 100 per cent oxygen If the decreased arterial oxygen saturation is due to impaired diffusion of oxygen in the lungs, complete oxygen saturation is obtained on breathing 100 per cent oxygen, and the oxygen tension rises to a practically normal level In the presence of a large right to left shunt, there is decreased oxygen saturation of the arterial blood even on breathing 100

pulse and piezoelectric arterial pulse curve. Six tracings can be recorded simultaneously. It is also coupled to a direct-writing electrocardiograph with jet recorders (Mingograf). The ECG, phonocardiogram, or pressure curves can be followed continuously on a cathode-ray oscilloscope.

Testing of the time lag between the different recordings showed that the pressure impulse occurred 0.01 sec later than the ECG and the phonocardiogram. This applied in pressure recordings with no. 6 to 10 catheters.

Blood-Gas Analyses

The analyses were earlier made exclusively with Van Slyke's apparatus (668). In recent years we have also used a photo-

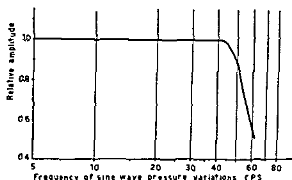


Fig. 115.—Frequency response curve of the strain gauge manometer (Elema) attached to a no. 8 Lehman catheter Damping varied from 0.7 to 0.8

electric method with reflected light, the hemoreflector designed by Brinkman (manufactured by Kipp & Zonen, Delft, Holland) The reliability of the method has been studied by Zijlstra (727) and Rodrigo (560). Only small quantities of blood are required (about 1 ml), and each determination, including taking the sample, can be performed in a couple of minutes This has considerable advantages in the investigation of congenital heart disease. In the presence of a shunt, there is incomplete mixing of the blood in the heart, particularly in the atria. Consequently, it is essential to take many samples from the same chamber. A method requiring large quantities of blood cannot therefore be used in small children. The loss of blood is too great and the oxygen capacity undergoes changes. This also applies to infants when a large number of blood samples are analyzed with the photoelectric method, and a small blood transfusion must then be given

It is of paramount importance that the determinations of oxygen saturation be ready before catheterization is finished, so that the results can be evaluated immediately. One reason for this is that, on the basis of these findings, one can decide whether an angiocardiographic examination should be performed and, if it is to be done, into what chamber or vessel the contrast medium is to be injected. Angiocardiography is carried out immediately after catheterization.

At each catheterization, one sample was always taken for analysis according to Van Slyke, for determination of the oxygen capacity of the blood. Double determinations of oxygen saturation were also made on each sample with the photoelectric method. The standard error of the method was calculated on 100 double determinations taken at random. The standard error of a single determination was found to be ±1.25 per cent, and the standard error of the mean of two determinations to be ± 0.89 per cent saturation.

In those cases in which the cardiac output and stroke volume are given, all blood-gas analyses were performed with the manome [illegible] lculating the [illegible] paci[illegible] tent of the blood sample, and vice versa, corrections were made for physically dissolved oxygen Since the oxygen capacity usually increases during exercise, the capacity was determined at each work load

Calculations of Blood Flow and Shunts

In heart disease associated with a shunt, calculation of the blood flow by the direct Fick method is subject to large sources of error It is often difficult to obtain mixed

venous blood, particularly in the presence of atrial septal defect or anomalous drainage of pulmonary veins. In patent ductus arteriosus, the blood in the pulmonary artery is not completely mixed. Moreover, considerable variations are often found in the oxygen saturation of blood from the same chamber of the heart from one moment to the next. This applies in particular to infants and in certain severe cardiac malformations. In transposition of the great vessels, the variations associated with respiration may be appreciable (see p. 787).

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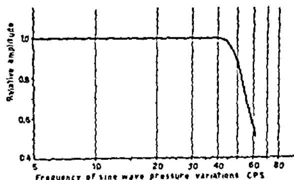


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In those cases in which the cardiac output and stroke volume are given, all blood-gas analyses were performed with the manometric Van Slyke apparatus In calculating the oxygen saturation from the oxygen capacity of hemoglobin and the oxygen content of the blood sample, and vice versa, corrections were made for physically dissolved oxygen Since the oxygen capacity usually increases during exercise, the capacity was determined at each work load

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In heart disease associated with a shunt, calculation of the blood flow by the direct Fick method is subject to large sources of error. It is often difficult to obtain mixed

through the leg vein and another through the arm vein.

In thoracic aortography, the catheter was inserted through the radial artery of the right arm. The incision in the artery was made about 2 cm distal to its origin from the brachial artery. It is important that the intima should not be rolled up when the catheter is inserted. On the advice of Dr. H. E. Hanson, two Deenatel sutures were made in the margin of the incision to avoid this occurrence. The opening in the vessel is widened by this means, and the intima is also secured by the sutures. After catheterization, the vessel was sutured, even though the risk of circulatory impairment after ligation of the radial artery is generally inappreciable.

Normal Values

The findings at cardiac catheterization in healthy subjects have been reported by several authors (42, 190, 199, 202, 260, 263, 345, 558, 610, 669). These series have, as a rule, been composed only of adults. We have therefore considered it of interest to report, in the following, our findings in a series of children investigated with the same technique as that used for studying the hemodynamics in our series of congenital heart disease.

We had the opportunity of performing cardiac catheterization in 30 individuals whom we regarded as healthy. Their ages ranged from 2½ to 19 years. They were sent to our department because heart disease was suspected. All of them were asymptomatic. In some cases a systolic murmur had been heard at a routine examination. In others, mass miniature roentgenologic examination had shown a wide "pulmonary arc." In all these cases, we regarded the murmur as physiologic, and the width of the main trunk of the pulmonary artery as within the normal range of variations. Five of the cases with an unusually wide pulmonary artery were not included among the normal cases. They have been reported under the heading of idiopathic dilatation of the pulmonary artery, even though—in our opinion—it was only a question of extreme normal variants in completely healthy individuals.

TABLE 1.—Normal Cases. Pressure, mm Hg, Determined at Right Cardiac Catheterization*

	RA			RV		PA			PCV		
	a	v	Mean	Syst	End-diast	Syst	Diast	Mean	a	v	Mean
n	30	30	30	30	30	29	23	30	15	15	22
mean	5 ± 0.4	4 ± 0.4	3 ± 0.4	23 ± 0.7	4 ± 0.5	19 ± 0.5	8 ± 0.4	13 ± 0.5	8 ± 0.5	11 ± 0.8	7 ± 0.5
σ	2.2	2.1	1.9	3.6	2.4	2.9	2.2	2.6	1.9	3.0	2.5
Range	0-9	0-8	0-6	14-32	0-10	14-27	2-12	8-19	5-11	6-18	2-11

*The following abbreviations are used in the tables: BrA, brachial artery; Fem A, femoral artery; IVC, inferior vena cava; LA, left atrium; LV, left ventricle; PA, pulmonary artery; PCV, pulmonary capillary venous pressure (171, 217); PV, pulmonary vein; RA, right atrium; RV, right ventricle; SVC, superior vena cava.

Nothing abnormal was found in the lungs of any of these children, and they were otherwise in good health.

Table 1 shows the results of the pressure [illegible] that is in relation to the body size, i.e., the anterior axillary line, which is easy to determine. In the normal cases, no correlation was present between the pulmonary artery pressure and age, but infants are lacking in this series. Systolic pressure in the right ventricle ranged from [illegible], but the pressure gradient never exceeded 8 mm Hg. The mean pressure in the pulmonary artery ranged from 8 to 19 mm Hg (mean 13 mm).

In pressure measurements in the pulmonary artery, the catheter lies in the direction of the blood flow. Owing to the pressure loss of velocity, the pressure recorded is lower than the actual pressure. If the width of the

per cent oxygen. If the shunt is small the saturation can, it is true, rise to 100 per cent, but the oxygen tension does not reach a normal value. In order to distinguish between an intracardiac and an intrapulmonary shunt, the pulmonary venous blood must be analyzed.

The systemic flow (Q_s) is calculated as follows.

$$Q_s \text{ (l/min)} = \frac{\dot{V}_{O_2}}{C_{O_2} \text{ (brach. art.)} - C_{O_2} \text{ (mixed venous blood)}} \quad (2)$$

Mixed venous blood is sampled from the pulmonary artery. If a left to right shunt is present, the mixed venous blood is sampled from the cavity directly proximal to the shunt, i.e., in atrial septal defect the blood is taken from the caval veins, in ventricular septal defect from the right atrium, and in patent ductus arteriosus from the right ventricle

The right to left shunt $= Q_s - Q_p$ (3)

The left to right shunt $= Q_p - Q_s$ (4)

If a mixed shunt is present, the right to left shunt is calculated as

$$R \rightarrow L \text{ shunt} = Q_s \cdot \frac{C_{O_2} \text{ (pulm. vein)} - C_{O_2} \text{ (brach art.)}}{C_{O_2} \text{ (pulm vein)} - C_{O_2} \text{ (mixed venous blood)}} \quad (5)$$

and the left to right shunt as.

$$L \rightarrow R \text{ shunt} = Q_p \cdot \frac{C_{O_2} \text{ (pulm art)} - C_{O_2} \text{ (mixed venous blood)}}{C_{O_2} \text{ (pulm vein)} - C_{O_2} \text{ (mixed venous blood)}} \quad (6)$$

The *vascular resistance* is most simply expressed as the ratio between the pressure gradient over the vascular bed and the blood flow This ratio is sometimes converted into absolute units, dyn sec cm^{-5} (289), but this does not imply any advantage. On the other hand, it is essential to put the cardiac output in relation to a body parameter It can be related to the body surface, the blood flow being expressed as the cardiac index (l/min/sq m of body surface area) A better body parameter is, however, the predicted basal oxygen uptake (346).

Insertion of the Catheter

The customary technique has been used. The median basilic vein of the left arm is the site of choice. In small children it is, however, often necessary to use the saphenous vein in the groin.

In adults a double-lumen catheter is routinely used, and sometimes another catheter is introduced into the right atrium, the percutaneous technique described by Seldinger (588) being used. With the same technique, a polyethylene tube is inserted into a systemic artery. A thin nylon cord was substituted for the spiral metal guide, permitting the use of considerably finer puncture needles. The technique could then be used even in infants. In many children, however, arterial puncture was unsuccessful.

For angiocardiography, a very wide catheter must be inserted. We now use catheters with a large lumen and a thin wall (Lehman), with several holes in the sides of the catheter, at most 1 cm from the tip This type of catheter does not recoil during rapid injection. It cannot, however, be used for pressure measurements in the wedge position. Therefore, we nowadays use different catheters for catheterization and for angiocardiography.

When the catheter is introduced into the leg, it passes easily into the left side of the heart if there is an atrial septal defect and strikingly often when the foramen ovale is patent. On the other hand, it is more difficult to advance the catheter into the pulmonary artery It is often necessary to place it in a loop in the right atrium. Great care is required to avoid the formation of knots. If the catheter is inserted into the arm, it is more difficult to pass through an atrial septal defect When this malformation was suspected, we introduced one catheter

Male, age 20 At rest Stroke volume = 132 ml
Ejection time = 0.32"

During exercise Stroke volume = 127 ml
Ejection time = 0.16"

Fig 117.—Effect of loss of velocity on the pressure recorded in the pulmonary ... exercise in a healthy man, aged 20 At rest, there is no pressu... and pulmonary artery. The stroke volume is as large during ... time is shortened by half, indicating that the velocity of flow ... pears and is most marked during the phase of rapid ejection.

pulmonary orifice is normal and the flow is *not increased*, the rate of flow is so low that the pressure loss of velocity need not be taken into account. This does not apply in pulmonary stenosis (p. 181) in the presence of a large flow due to a left to right shunt (Fig. 116) or during exercise (364) (see Fig. 117).

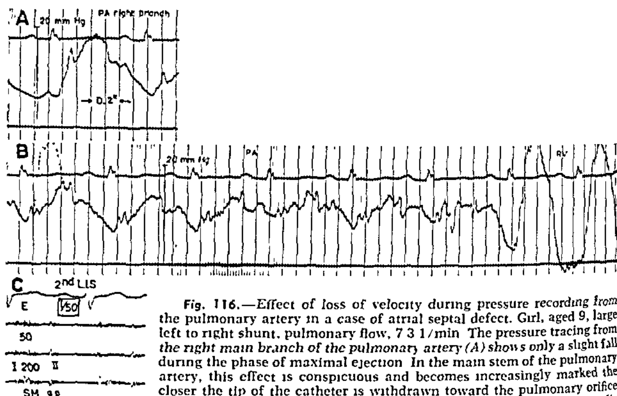


Fig. 116.—Effect of loss of velocity during pressure recording from the pulmonary artery in a case of atrial septal defect. Girl, aged 9, large left to right shunt, pulmonary flow, 7.3 l/min. The pressure tracing from *the right main branch of the pulmonary artery (A) shows* only a slight fall during the phase of maximal ejection. In the main stem of the pulmonary artery, this effect is conspicuous and becomes increasingly marked the closer the tip of the catheter is withdrawn toward the pulmonary orifice *(B)*. The fall in pressure coincides with the maximum of the systolic murmur *(C)*. Dotted line denotes the shape of the tracing in the state of normal flow. Assuming the diameter of the pulmonary orifice to be 1.5 cm and emptying of the greater part of the right ventricle to take place within 0.15 sec, the calculated value of pressure loss of velocity will be 18 mm Hg. Since values for width of the orifice and ejection time of the ventricle are most uncertain, only an approximate value will be obtained for pressure loss of velocity. This value was, *however, of the same order of magnitude as the pressure gradient between the right* ventricle and pulmonary artery. Systolic pressure in right ventricle, 25 mm Hg; in pulmonary artery valve region, 13 mm Hg. Boxed figure denotes degree of amplification, other figures denote standard frequency of the filters. *I*, 1st sound, *II*, 2nd sound, *SM*, systolic murmur, *a* and *p*, aortic and pulmonary components of the 2nd sound, *LIS*, left interspace. ECG in *A–B* = lead II, in *C* = lead I.

The end-diastolic pressure in the right ventricle was in fairly good correlation with *the right atrial systolic pressure. The latter* mean pressure ranged from 0 to 6 mm Hg (mean 3 mm). A characteristic feature of the pressure curve from the right atrium is that the *a* wave is generally higher than the *v* wave. In one case, it was possible to pass through a patent foramen ovale, the pressure in the left atrium was higher than that in the right, and the *v* wave was higher than the *a* wave. This is also apparent from the pulmonary capillary venous pressure. This matter will be discussed further in connection with the hemodynamics in atrial septal defect (p. 437).

During the pressure recording, the catheter must lie freely in the relevant chamber of the heart or in the vessel. Examples of *the effect of some artefacts* are shown in Figure 118. If the catheter lies in the auricular appendage, high atrial pressure and an abnormally shaped curve are obtained. This also applies if the tip is in contact with one of the atrioventricular valves or if the catheter moves between the pulmonary artery

(in systole) and the right ventricle (in diastole). When the catheter is introduced [illegible] atrial

valve It is easier to place the catheter freely in the left atrium if it is introduced through *a leg vein* If the catheter lies in a coronary vein, a tracing is obtained which often resembles a ventricular tracing. If its *position cannot* be established at fluoroscopy, it can easily be ascertained by means of a blood determination, since the oxygen saturation is low in the coronary sinus.

In order to establish the presence of a shunt, the normal difference between the oxygen saturation of the different vessels and chambers of the heart must be known These values are assembled in Table 2.

Normally, small variations are present between the oxygen saturation of the right side of the heart, the venae cavae, and the pulmonary artery. The difference between the inferior and the superior vena cava is highly significant The higher oxygen saturation of the inferior vena cava is to be ascribed to the low arteriovenous oxygen difference in the renal circulation Owing to the laminary flow, the blood is incompletely mixed in the veins This is particularly apparent in the inferior vena cava, since some of its blood comes *from the kidneys* and has a high oxygen saturation, and some from the liver with a low oxygen saturation This may explain the high *standard* error of the difference in oxygen saturation of the blood from the superior and inferior vena cava In order to demonstrate *the deficient* mixing, two samples were taken in immediate sequence from the inferior vena cava, the catheter being rotated before taking the second sample. This series comprised 31 cases of various congenital cardiac defects. The standard deviation of the difference between the two samples is 5.9 per cent saturation (at a mean saturation of 68 per cent). Presumably, the variations are somewhat greater in these pathologic cases than in a normal series

The oxygen saturation of the blood in the right atrium should be lower than that of the mixed caval blood, since the former also receives blood with a very low oxygen saturation from the coronary sinus. In our series the difference is, however, only probably significant. The blood in the right ventricle was somewhat less saturated than that in the right atrium, and the difference is highly significant This is to be ascribed to the fact that part of the blood from the coronary sinus is not mixed with that in the atrium, but flows directly into the ventricle. We also found a highly significant difference between the mixed caval blood and the blood from the pulmonary artery, in which mixing was effective No significant difference was present between samples from the right ventricle and the pulmonary artery, thus indicating effective mixing already in *the ventricle.* This does not apply in the presence of a large left to right shunt.

Nor was there any signficant difference between the samples from the superior vena cava and those from the pulmonary artery *The higher* oxygen content of the blood from the inferior vena cava seems to be reduced by mixing with the blood of a lower oxygen content from the coronary sinus The blood from the superior vena cava is thus representative of *mixed venous blood*

TABLE 2.—THIRTY NORMAL CASES· DIFFERENCE BETWEEN PERCENTAGE OXYGEN SATURATION OF BLOOD SAMPLES FROM DIFFERENT CHAMBERS OF THE HEART AND VESSELS*†

	IVC — SVC	$\frac{IVC + SVC}{2}$ — RA	$\frac{IVC + SVC}{2}$ — PA	RA — PA	RV — PA	SVC — PA
mean	+ 5 6 ± 0 84	+ 0 9 ± 0 43	+ 2 5 ± 0 34	+ 1 4 ± 0 37	0 0 ± 0.36	− 0 2 ± 0 56
σ	4 59	2 37	1 88	2 03	1 97	3 08
P	< 0 001	0 05–0 02	< 0 001	< 0 001	> 0 9	0.7–0 6

*Oxygen capacity varied between 14 5 and 20 4 vol % (mean 17 1)
†For abbreviations see Table 1, p 119

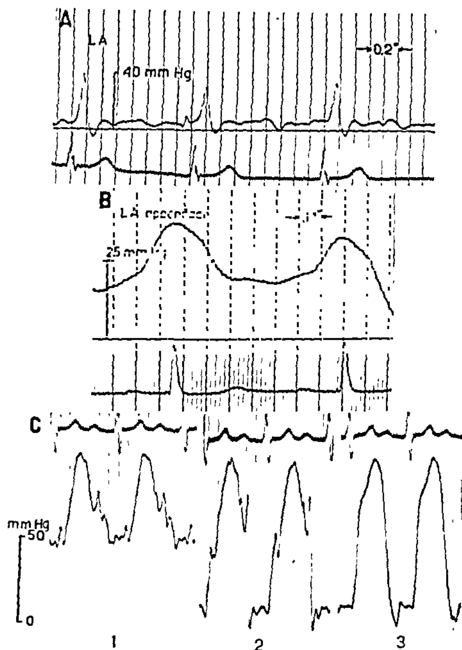


Fig. 118.—Artefacts in pressure recording *A*, when catheter tip in left atrium is in contact with mitral valve, rapid rise in pressure is obtained during isometric phase of ventricular contraction Amplitude varies. *B*, if catheter tip lies in auricular appendage, an atypical atrial curve is obtained Opening of catheter lies in contact with atrial wall. Similar curves can be obtained under these conditions from right atrium *C*, catheter tip (*1*) in main trunk of pulmonary artery and (*3*) in right ventricle. The deviating type of curve in (*2*) was caused by the catheter's having moved between the pulmonary artery (in systole) and the right ventricle (in diastole), owing to the ventricular contractions This occurs not uncommonly in mild valvular pulmonary stenosis, and the withdrawal curve can then be confused with that obtained in infundibular stenosis (cf. Fig. 178, p 183)

trast medium is so weak that it is deflected by the intracardiac blood flows in its path. We have hitherto seen no endocardial damage that could be ascribed to the jet or to the impact of the catheter.

In order to visualize an atrial septal defect with a left to right shunt, it has been found necessary to inject the contrast medium into the left atrium The injection must take place very rapidly, i e, within ½ to 1 sec Consequently, only large-bore catheters can be used Owing to the thickness of the catheter, it can as a rule be inserted only through the saphenous vein However, this route invariably affords the best possibility of passing through an atrial septal defect or a patent foramen ovale

Angiocardiography is generally performed immediately after cardiac catheterization The patient lies on a specially constructed table with a movable top (Fig. 119). After the position of the catheter tip has been carefully checked and the catheter secured, the top is slid forward over the apparatus for angiocardiography (Figs. 120 and 121).

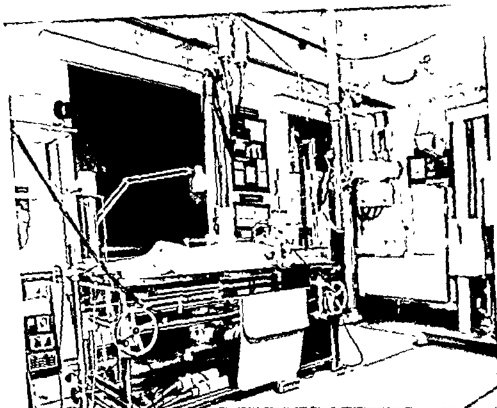


Fig. 119.—View of the heart laboratory and equipment used for cardiac catheterization and angiocardiography

Injection of contrast medium into the right ventricle is unquestionably the most common procedure. It is then of paramount importance to avoid injection into a coronary vein. It may sometimes be difficult at fluoroscopy to determine whether the catheter tip lies in the infundibulum of the right ventricle or in a coronary vein. In order to ascertain the position of the catheter *with absolute certainty*, we have used the following technique. The catheter is advanced into the pulmonary artery or possibly into the aorta The patient's arms are stretched above his head. The catheter is then with-

This nevertheless applies only in normal subjects at rest. *During exercise, the conditions undergo a considerable change.* It is also likely that it does not invariably apply at rest in the presence of heart disease. This *is because a change in renal blood flow* alters the difference between the oxygen saturation of blood from the two caval veins. Consequently, it seems most correct, *in cases with a left to right shunt,* to use the mean values for the superior and inferior vena cava for determinations of mixed venous blood (343). It might, on first thoughts, seem incorrect to use the mean value, since the flow through the latter is greater than that through the former. However, in small children, the blood flow through the upper part of the body is relatively greater than in adults Consequently, the calculated value for the mixed caval blood probably differs only inappreciably from the true value.

With our technique, the normal arterial oxygen saturation was found to be 97 per cent (range 94 to 98 per cent).

ANGIOCARDIOGRAPHY

The great value of angiocardiography in *the diagnosis of congenital heart disease* can no longer be doubted. Nevertheless, in view of the risks still associated with it, *angiocardiography should not be regarded as a routine method to be used indiscriminately.* It should be reserved for specific problems and for the anatomic localization of the affected parts of the heart or blood vessels.

Angiocardiography often provides the final considerations needed by the surgeon to assess the operability and the appropriate surgical technique. The examination should therefore be planned with a view to giving the surgeon an exact and complete answer to all the questions of consequence for the operation. These cover such matters as the site and extent of stenosis, the size and localization of a septal defect, the degree of over-riding of the aorta and the appearance of the systemic arteries and pulmonary vessels. In order to reach this goal, we have found it expedient to adopt *two procedures. One is to inject* the contrast medium as closely as possible into the area involved, i.e., *selective angiocardiography* The other is to use a technique permitting *an extremely rapid series of exposures.* The importance of the latter factor has been strongly emphasized by Wegelius and Lind (689).

Selective angiocardiography. — This procedure, introduced by Chavez *et al* (153) and elaborated by Jönsson *et al* (367, 369) and usually performed by injection of the contrast medium through a catheter, was used in 628 of our cases Venous angiocardiography was performed in 13 cases only.

The *catheters* first used were of the Cournand type (manufactured by the U.S. Catheter & Instrument Co., Glens Falls, N.Y.) The manufacturers subsequently modified *the construction by placing a joint of softer* and less resistant material between the metal cone-lug and the actual catheter. This made them less suitable for our technique. In order to introduce the necessary *quantity* of contrast medium per unit time, the injection must be made, in the majority of cases, under high pressure. As a result, the *soft joint of the catheters in question* is apt to burst. In order to reduce the pressure of injection, but to retain or possibly increase *the quantity injected per unit time,* we have *tested a new type of catheter, the* Lehman catheter (manufactured by the same firm) Its lumen is considerably increased at the cost of the thickness of the wall, whereas *the outer diameter is unchanged.* This type of catheter has proved to be superior in many respects and we now use it almost exclusively To avoid recoil of the catheter tip, and to achieve better mixing of the blood and contrast medium, we have for about the past two years used, almost without exception, such catheters with holes bored in the side walls, the hole in the tip being sealed. The catheter manufactured by Rusch in Germany has also been tested, but has proved to be unsuitable for our examinations

Even in rapid injection, the jet of con-

trast medium is so weak that it is deflected by the intracardiac blood flows in its path. We have hitherto seen no endocardial damage that could be ascribed to the jet or to the impact of the catheter.

In order to visualize an atrial septal defect with a left to right shunt, it has been found necessary to inject the contrast medium into the left atrium The injection constructed table with a movable top (Fig. 119). After the position of the catheter tip has been carefully checked and the catheter secured, the top is slid forward over the apparatus for angiocardiography (Figs. 120 and 121).

Injection of contrast medium into the right ventricle is unquestionably the most common procedure It is then of paramount

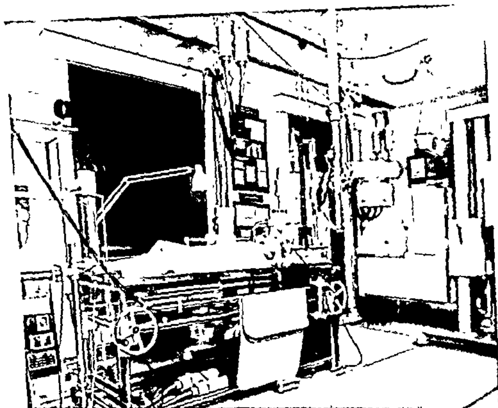


Fig 119.—View of the heart laboratory and equipment used for cardiac catheterization and angiocardiography

must take place very rapidly, i.e , within ½ to 1 sec Consequently, only large-bore catheters can be used Owing to the thickness of the catheter, it can as a rule be inserted only through the saphenous vein. However, this route invariably affords the best possibility of passing through an atrial septal defect or a patent foramen ovale.

Angiocardiography is generally performed immediately after cardiac catheterization The patient lies on a specially

importance to avoid injection into a coronary vein. It may sometimes be difficult at fluoroscopy to determine whether the catheter tip lies in the infundibulum of the right ventricle or in a coronary vein In order to ascertain the position of the catheter *with absolute certainty*, we have used the following technique The catheter is advanced into the pulmonary artery or possibly into the aorta. The patient's arms are stretched above his head. The catheter is then with-

This nevertheless applies only in normal subjects at rest. During exercise, the conditions undergo a considerable change. It is also likely that it does not invariably apply at rest in the presence of heart disease. This is because a change in renal blood flow *alters the difference between the oxygen* saturation of blood from the two caval veins. Consequently, it seems most correct, in cases with a left to right shunt, to use the mean values for the superior and inferior vena cava for determinations of mixed venous blood (343). It might, on first thoughts, seem incorrect to use the mean value, since the flow through the latter is greater than that through the former. However, in small children, the blood flow through the upper part of the body is relatively greater than in adults. Consequently, *the calculated value for the mixed caval* blood probably differs only inappreciably from the true value.

With our technique, the normal arterial oxygen saturation was found to be 97 per cent (range 94 to 98 per cent).

ANGIOCARDIOGRAPHY

The great value of angiocardiography in the diagnosis of congenital heart disease can no longer be doubted Nevertheless, in view of the risks still associated with it, angiocardiography should not be regarded as a routine method to be used indiscriminately It should be reserved for specific problems and for the anatomic localization of the affected parts of the heart or blood vessels.

Angiocardiography often provides the *final considerations needed by the surgeon* to assess the operability and the appropriate surgical technique The examination should therefore be planned with a view to giving the surgeon an exact and complete answer to all the questions of consequence for the operation. These cover such matters as the site and extent of *stenosis, the size* and localization of a septal defect, the degree of over-riding of the aorta and the appearance of the systemic arteries and pulmonary vessels. In order to reach this goal, we have found it expedient to adopt two procedures. One is to inject the contrast medium as closely as possible into the area involved, i e., *selective angiocardiography* The other is to use a technique permitting an extremely rapid series of exposures The *importance of the latter factor has been* strongly emphasized by Wegelius and Lind (689).

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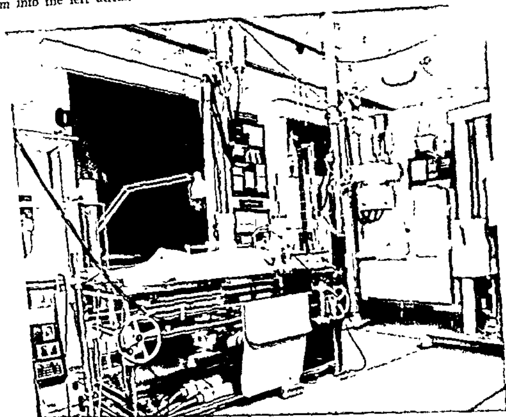
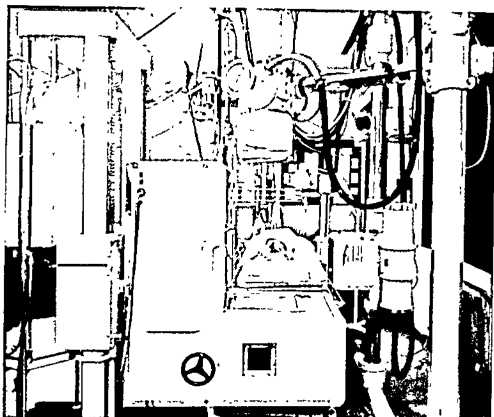


Fig 119 —View of the heart laboratory and equipment used for cardiac catheterization and angiocardiography

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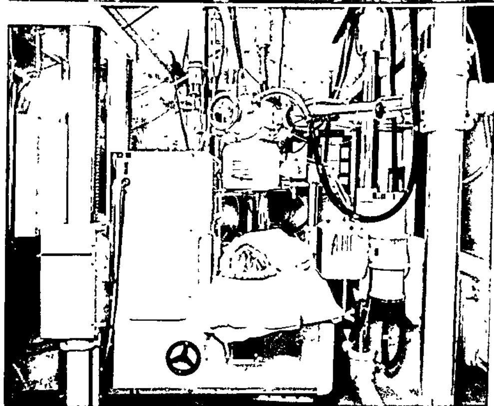


Fig. 120 (*above*).—Serial film changer, table top in position for catheterization

Fig. 121 (*below*).—Table top over the serial film changer and in position for angiocardiography.

rawn into the right ventricle under fluoro-copic control and eventually continuous ressure recording. If the catheter cannot e advanced into the pulmonary artery or he aorta, its position can be ascertained by visualized. If, on the contrary, the tip of the catheter is in the ventricle, the contrast medium spreads diffusely and disappears rapidly.

When the tip has entered the ventricle, it

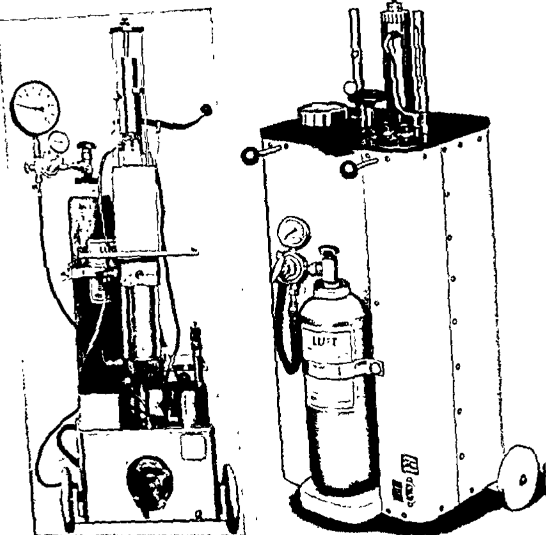


Fig 122.—Automatic syringe used for angiocardiography *Left*, prototype, *right*, modern style

pressure recording, determination of the oxygen saturation, and the manual injection of a small quantity of contrast medium under fluoroscopic control If the catheter lies in a coronary vein, a damped ventricular curve is obtained, oxygen saturation is extremely low, and the coronary vein is

is important for the catheter to be placed in a wide loop, supported lightly by the atrial wall This decreases the risk of the tip's being repelled into the atrium by the pressure of injection The tip of the catheter should be in the inflow tract, if possible in the apex, and not in the infundibulum

SELECTIVE ANGIOCARDIOGRAPHY OF THE LEFT SIDE OF THE HEART.—With the advances in modern heart surgery, increasingly great demands have been made on preoperative special diagnosis of mitral and aortic disease. Of the techniques used for this purpose, angiocardiography has proved to be of particular value, since it permits exact anatomic localization of the patho-

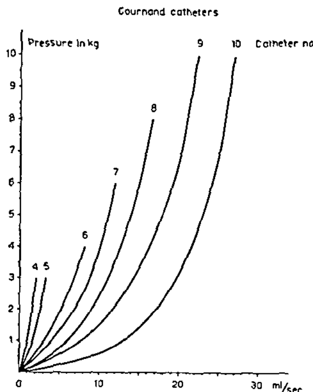


Fig. 123—Chart showing quantity of contrast medium per unit time for Cournand catheters at various pressures Chart refers only to curves produced by injector shown in Figure 122 (*left*).

logic changes. An acceptable picture of the nature and extent of the process has, however, been obtained only by injection of the contrast medium directly into the left side of the heart. In mitral disease, this has been done by transthoracic puncture of the left atrium, in aortic disease and as a complementary examination in some cases of mitral disease, transthoracic puncture of the left ventricle is used (168, 421, 540)

THORACIC AORTOGRAPHY.—This was carried out in two different ways. In some cases it was performed according to the technique of Jönsson *et al.* (370), with exposure of the radial artery on the right side and insertion of the catheter into it. In others, Seldinger's method (588) was used, with percutaneous insertion of a polyethylene catheter through the brachial artery. However, the latter method cannot as a rule be used in children under 4 to 5 years old, but it has considerable advantages in several respects.

The contrast medium is injected with an *automatic syringe* on Gidlund's principle (Fig. 122). It is provided with two barrels, one for the contrast medium and one for compressed air. The latter is used to push the piston down into the barrel containing contrast medium. The construction of the syringe is such that the risk of air embolism is entirely eliminated The contrast medium is warmed to blood heat. The pressure barrel is connected to a cylinder containing compressed air. The desired pressure is obtained by means of a reducing valve. The syringe is linked to the film changer and starts it at the required time.

In order to obtain satisfactory visualization, the contrast medium must be injected rapidly. into the right or the left ventricle or the pulmonary artery within 2 to 3 sec, and into the aorta within 2 sec These figures apply with a pulse rate of 60 per minute (Figs. 123 and 124). With a rising pulse rate, the pressure of injection is correspondingly increased, as shown in Figure 125. Injection into the left atrium, for a study of atrial septal defect, must be made still more rapidly It must not take more than ½ to 1 sec, and a frequent change of exposures must be made during this time

We have made repeated tests with injection of the contrast medium with a manually operated syringe. The results have been poor in the majority of cases In our opinion, the automatic syringe is invaluable in the examination of children

As *contrast medium,* we have used a 70 per cent solution of the perabrodil-Diodrast group or of Urokon. Urokon was found to be superior to the other media, with regard both to density and to the absence of side-effects The quantity was 1 2 cc per kilo-

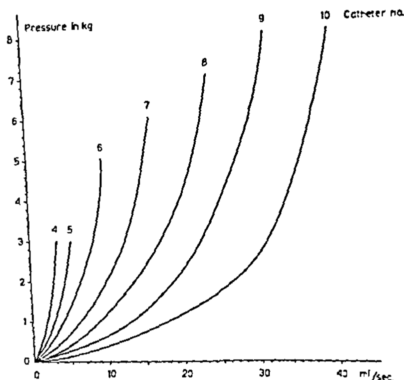


Fig. 124.—Chart showing quantity of contrast medium per unit time for Lehman catheters at various pressures. Chart refers only to curves produced by injector shown in Figure 122 (*left*).

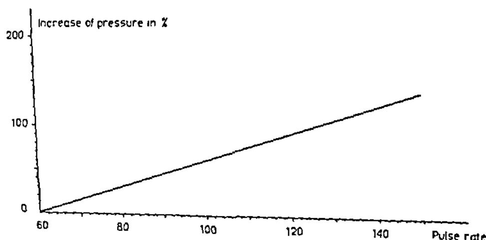


Fig. 125.—Pressure corrections required for different pulse rates.

gram of body weight, except in left-sided angiocardiography and thoracic angiocardiography, when it was decreased to 0.8-1 cc per kilogram, and in cases of a large left to right shunt, when it was sometimes increased to 1.4 cc per kilogram. We found in animal experiments that a further increase in the amount of contrast medium was associated with considerably greater risks. In the dog, injection of a dose of 70 per cent perabrodil-Diodrast in excess of 2 cc per kilogram of body weight resulted in death during the investigation. Recently we have used Urografin, which is even less toxic.

We always perform angiocardiography under general anesthesia, with administration of oxygen.

After thorough study, we have come to the conclusion that the information required by the surgeon is usually best provided by *frontal* and *lateral projections* An exception must be made for atrial septal defect and mitral and aortic valvular disease, when *oblique projections* are used.

In order to obtain the right quality, it has been found advantageous to use the same *current intensity* for both the frontal and the lateral projection. We have consistently used a current of 350 ma for each tube, i.e., a total of 700 ma, and kilovoltage ranging from 70 to 130 All exposures have been taken with a three-phase apparatus, equipped with a contactor with thyratrone tubes.

Our *roentgen apparatus*, manufactured by the Elema-Schonander Co , Stockholm, has a capacity of 1,000 ma at 125 kv or of 700 ma at 150 kv. Cross-grids (constructed by the Elema-Schonander Co , Stockholm) are used in order to decrease the appreciable scattered radiation which arises on simultaneous exposure with two tubes.

The *angiocardiographic apparatus* is a roll-film serial changer taking films in two planes at right angles to each other (Figs 120 and 121). The serial changer is constructed by Elema-Schonander Co., Stockholm, according to Gidlund's design. The maximum size of each picture is 30 × 30 cm (12 × 12 in.), and each roll of film is 25 meters (83 ft) long. The exposed strip of film is cut off and removed for developing with a simple manual maneuver. The unexposed film is left in the serial changer for subsequent investigations. By means of a program selector, the rate of exposure can be varied from 12 exposures per second to 1 every 10 sec. In addition, it is possible to vary the rate of exposure in the course of the investigation.

Siemens' *roentgen tubes*, which in series of at least 100 frames tolerate 150 kv at 350 ma, are used. The exposures are made simultaneously with both tubes. We have used an *exposure time* of 0.003 sec for children under 3 to 4 years old, 0.01 to 0.02 sec for older children, and 0 03 to 0.045 sec for adults.

The *electrocardiogram* is usually recorded during the angiocardiographic examination. An *exposure marking* on the ECG is then obtained simultaneously. By this means it is possible not only to determine the exact position of each exposure in the cardiac cycle, but also to keep a check on the heart rhythm during the actual injection of contrast medium and to control the function of the apparatus Ventricular extrasystoles usually appear during the actual injection, especially when made in the right ventricle, but cease once it is ended

ANESTHESIA IN CARDIAC CATHETERIZATION AND ANGIOCARDIOGRAPHY

By Olof Norlander, M.D.*

During cardiac catheterization, the patient should be calm, relaxed, and completely at rest In older children, a light sedative usually suffices to produce this state. In children under 4 to 5 years old, it is generally necessary to resort to basal anesthesia. It is true that every form of narcosis alters the hemodynamic conditions, but it is of still greater consequence for the interpretation of the results for the patient to be in the same state throughout the investigation

During angiocardiographic examination, surgical anesthesia is required, owing to

*Head of the Department of Anesthesiology, Thoracic Clinics, Karolinska Sjukhuset

the pain and reflexes elicited by the contrast medium.

During recent years, we have used the following procedure.

ADULTS AND CHILDREN OVER 4 TO 5 YEARS OLD.—The patients are prepared as for any form of planned anesthesia; i.e., no food or drink is given for the preceding 12 hours. One to 2 hours before the examination is started, a suitable barbiturate is given by mouth or injected subcutaneously as a sedative. One to 1½ hours before angiocardiography, additional premedication is given in the form of morphine-scopolamine or Phenergan, according to age.

CHILDREN UNDER 4 TO 5 YEARS OLD.—These patients are usually given rectal basal anesthesia with tribromoethanol (Avertin). The rectal catheter remains in situ during the investigation, so that an additional dose may be administered if required. In order to hasten resorption, the anesthetic is dissolved in a small quantity of surface-tension-decreasing medium (benzalkonium chloride). The dosage is calculated on the basis of the body weight, the dose being kept on the low side.

PROCEDURE IN ANGIOCARDIOGRAPHY.—The object of anesthesia is to secure *freedom from pain* for the patient, *optimal oxygen saturation of the tissues*, and the *best possible working conditions for the roentgenologist*. In our opinion, these conditions are best fulfilled in children by means of *adequate basal anesthesia, combined with* a sufficient dose of succinylcholine to produce complete muscular paralysis. In adults, as well as in cases in which basal anesthesia is inadequate, a small dose of barbiturate is injected, preferably into a peripheral vein. If this is not feasible, a dilute (0.5-0.75 per cent) solution of the barbiturate is injected into the heart catheter. The injection is made extremely slowly with a few pauses. When the patient is well asleep, but not too deeply, succinylcholine is given in a quantity of about 1 mg per kilogram of body weight. As soon as muscular paralysis is produced, artificial respiration is started with a balloon and a well-fitting face mask in a semiclosed to-and-fro system without soda lime canister. 100 per cent oxygen or a mixture of maximally 50 per cent nitrous oxide in oxygen at high flow rates is used.

Angiocardiography is performed with the patient in apnea and the lungs inflated to an intrapulmonary pressure of +5 to +20 cm of water. In this way, it is possible to obtain good filling of the right side of the heart and generally of the left side as well. This is because there is less dilution of the contrast medium in the pulmonary circulation because of the partial decrease in blood flow through the lungs at increased intrapulmonary pressures. The period of increased intrapulmonary pressure should be of short duration, since a serious drop in systemic blood pressure may otherwise occur.

SELECTIVE ANGIOCARDIOGRAPHY OF THE LEFT SIDE OF THE HEART.—This procedure involves direct puncture of the left atrium or the left ventricle in patients with mitral or aortic valvular disease, who are poor anesthetic risks. Consequently, special precautions are taken; e.g., in most cases we use an intratracheal tube connected to a semiclosed circle-circuit anesthesia apparatus. Before and during injection of contrast medium through the heart needle, the patient is kept in light plane anesthesia with a combination of barbiturate and oxygen or a mixture of 50 per cent nitrous oxide in oxygen. Muscular paralysis is produced by succinylcholine before the skin is punctured with the heart needle. The intratracheal tube is not removed until the patient is completely awake and there are no signs of complications, as, for instance, pneumothorax. During the whole procedure, the ECG and systemic blood pressure are recorded continuously and observed on the [illegible]

[illegible], as complications may occur.

gram of body weight, except in left-sided angiocardiography and thoracic angiocardiography, when it was decreased to 0.8-1 cc per kilogram, and in cases of a large left to right shunt, when it was sometimes increased to 1.4 cc per kilogram. We found in animal experiments that a further increase in the amount of contrast medium was associated with considerably greater risks. In the dog, injection of a dose of 70 per cent perabrodil-Diodrast in excess of 2 cc per kilogram of body weight resulted in death during the investigation. Recently we have used Urografin, which is even less toxic.

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connection with a repeated injection of contrast medium At the first injection, the angiocardiographic apparatus had been wrongly connected. Deaths had been reported previously in association with repeated injections at close intervals (652).

2. A 7-year-old girl (large ventricular septal defect with a mixed shunt) died 10 minutes after the injection of contrast medium This was one of our first angiocardiographic examinations, and the syringe then used was so constructed that a small air bubble in the connecting tube could not be ruled out with certainty. No air embolism was found at autopsy, but neither could any other damage be demonstrated

3 A 3-year-old girl (persistent ostium atrioventriculare commune) died 10 minutes after the injection of contrast medium. Before the examination she was in extremely poor general condition and cardiac failure was present

4 A 3-year-old girl (ventricular septal defect and ECG changes suggestive of endocardial fibroelastosis), examined under general anesthesia, did not wake from the anesthesia High fever appeared, and she died one and one-half days after examination Autopsy was not performed, and the cause of death was uncertain The case has been classified under uncertain diagnosis.

5 A woman with primary pulmonary hypertension (not included in our series). cardiac arrest occurred five minutes after the injection of contrast medium

In addition, one infant (tetralogy of Fallot and pulmonary atresia) died soon after the induction of anesthesia, before the contrast medium had been injected.

Three of the deaths occurred in the beginning of the series On their account, the technique was changed. We now use a syringe which cannot give rise to air embolisms We do not perform angiocardiographic examination without the assistance of an anesthetist Repeated injections are never made in direct sequence.

In two cases of tetralogy of Fallot, spastic hemiparesis appeared in direct association with catheterization In one case it lasted only for an hour, but in the other complete restitution did not occur until several months later.

Occasional extrasystoles occurred in almost every case when the catheter passed through the infundibulum of the right ventricle. Other arrhythmias which appeared were a bundle-branch block in six cases, complete atrioventricular block in five, supraventricular tachycardia in four, auricular flutter in two, and ventricular tachycardia in five cases. All were of brief duration, except in one case of ventricular tachycardia in which death ensued (mentioned earlier) and one of A-V block, in which the arrhythmia persisted for a few days

A brief febrile reaction was observed in 22 cases In two, the catheter became tied in a knot in the right atrium. Both patients were infants. A soft catheter (no. 5) was inserted through the saphenous vein, and in order to pass it into the right ventricle, it [illegible] that vein and removed by laparotomy. Both patients survived, and one was successfully operated on later for the cardiac malformation

Venospasm was very seldom troublesome. To avoid it, it is essential that the patient is calm and that the catheter is not too thick in relation to the vein Only in one case (a 1-year-old patient) were we unsuccessful in introducing the catheter into the heart, two years later a further attempt was made and catheterization could be completed. In one case severe arteriospasm occurred on passage of the catheter through the brachial artery, with the object of performing thoracic aortography. After the examination, a severe circulatory disturbance was present in the arm, but complete restitution took place some months later.

Before angiocardiography, a small test dose of the contrast medium is injected. One patient showed hypersensitivity to it, and the examination was therefore not carried out.

4 Complications in Catheterization and Angiocardiography

A SURVEY of the risks and complications in these investigations has been published by the Committee on Cardiac Catheterization and Angiocardiography of the American Heart Association (164), as well as one from all Swedish laboratories (31).

Cardiac arrhythmias are very common in cardiac catheterization. Ventricular extrasystoles almost invariably appear on passage of the catheter through the infundibulum of the right ventricle. All types of arrhythmia have been described (406), but severe forms are rare. Other complications are uncommon Isolated cases have been reported of endocardial damage, knotting of the catheter, rupture of a coronary vein, and venous thrombosis. In pulmonary stenosis, the pulmonary flow may be entirely stopped on passage of the catheter through the orifice Even small air emboli may have serious consequences in patients with a right to left shunt, when the air bubbles may pass directly from the right side of the heart into the systemic circulation and enter the cerebral vessels. Consequently, the technique must be such that the occurrence of air embolisms can be ruled out with certainty In the presence of a right to left shunt it is also particularly important to observe strict asepsis, in view of the risk of cerebral abscess. Chills and fever of brief duration may occur during the first 24 hours after catheterization, due to pyrogenic substances in the catheter or the infusion apparatus. Furthermore, the possibility of brain damage in connection with angiocardiography must always be borne in mind, although the risk is fairly slight. This problem has been studied especially by Melin (485) and Kirstein *et al* (392).

The incidence of serious complications and deaths in association with cardiac catheterization and angiocardiography is dependent on how severely ill patients are examined If the patient is in good general condition, these investigations involve only an inappreciable risk. In patients with severe malformations and those in cardiac failure, the risk is greater It is, however, difficult to determine whether the examination has been the cause of death. Two of our patients, for example, died on the day on which catheterization had been planned to take place

In 837 cardiac catheterizations we had one death The patient, an 8-year-old girl with tetralogy of Fallot, had severe pulmonary stenosis and was in very poor general condition Ventricular tachycardia appeared during catheterization. At that time, our laboratory was not equipped with an oscilloscope, and the arrhythmia was not observed immediately The tachycardia could not be arrested, and the patient died 24 hours later.

In 728 angiocardiographic examinations, the following deaths occurred

1. An infant (mitral atresia) died in

	No. of Cases
Vascular ring	3
Valvular and subvalvular aortic stenosis	23 (+ 2)
Pulmonary incompetence	2
Ebstein's malformation of the tricuspid valve	6 (+ 1)
Tricuspid atresia	14
Tricuspid stenosis	4
Underdeveloped right ventricle without tricuspid atresia or stenosis	1
Mitral atresia	7
Mitral stenosis	2
Mitral incompetence	3
Transposition of the great vessels	20
Corrected transposition of the great vessels	3
Primary pulmonary hypertension	3
"Idiopathic" dilatation of the pulmonary artery	5
Uncertain diagnosis	4
Total	668 (+ 74)

Embryopathy

In 10 of our 668 cases, the mother had contracted a virus infection during the first months of pregnancy: in nine, the infection had been rubeola, and in one, mumps.

Heredity

A certain familial predisposition to congenital heart disease was demonstrated by McKeown *et al.* (455) but could not be shown in the series studied by Richards *et al* (577). In our series, we had two pairs of twins with patent ductus arteriosus, as well as one patient whose sister had earlier been operated on for this malformation In one case of coarctation of the aorta, another member of the family had the same cardiac anomaly. The series also includes two siblings with tetralogy of Fallot. An additional four patients each had a sibling with congenital heart disease, but of a type different from that of the patient in question

5 Case Material

OUR case material consists mainly of children. Most of them have been referred to us from other pediatric clinics in various parts of Sweden. Only a minor proportion come from Stockholm and a few from other countries. Operable cases of heart disease are over-represented among those sent to us from other hospitals. Severe malformations which are incompatible with survival for more than a few months are definitely under-represented, since such cases come only from the districts of the City and County of Stockholm, from which admissions are made. Consequently, it is impossible on the basis of our series to make any closer analysis of the actual incidence of various types of congenital heart disease in Sweden.

A few patients were sent to us only for roentgenologic examination, especially angiocardiography. They were adult patients from medical clinics, where a complete investigation, including cardiac catheterization, had been made. These cases will be discussed only with respect to the roentgenologic features.

In addition, a few adult patients, investigated at the Department of Clinical Physiology (director, Prof. T. Sjöstrand), are described, particularly with reference to the hemodynamics during exercise. These patients are not included in our case material.

It is not possible to make a consistent classification of congenital heart disease on the basis of the presence of shunts and their direction. We have instead classified our cases according to the anatomy of the malformations. Combined anomalies were often present; these cases have been accounted for under the malformation that we have considered to be the most important from the functional viewpoint. This will be discussed in more detail in the relevant chapters.

Our series is classified into the following groups. (The number of cases in which only a roentgenologic examination was made is given in brackets.)

	No. of Cases
Pulmonary stenosis with normal aortic root	70 (+14)
Pulmonary atresia with intact ventricular septum	2
Stenosis or atresia of pulmonary artery branches	2
Pulmonary stenosis with ventricular septal defect and right to left shunt (tetralogy of Fallot)	63 (+ 1)
Persistent truncus arteriosus	1 (+ 1)
Ventricular septal defect	117 (+ 5)
Ventricular septal defect with tricuspid incompetence	1
Defect involving the entire ventricular septum (single ventricle)	2
Atrial septal defect	77 (+ 7)
Persistent ostium atrioventriculare commune	7
Communication between left atrium and coronary sinus	2
Anomalous drainage of pulmonary veins	16 (+ 5)
Patent ductus arteriosus	138 (+ 4)
Aortic sinus aneurysm with communication to the right ventricle	2
Coarctation of the aorta	68 (+34)

	No. of Cases
Vascular ring	3
Valvular and subvalvular aortic stenosis	23 (+ 2)
Pulmonary incompetence	2
Ebstein's malformation of the tricuspid valve	6 (+ 1)
Tricuspid atresia	14
Tricuspid stenosis	4
Underdeveloped right ventricle without tricuspid atresia or stenosis	1
Mitral atresia	7
Mitral stenosis	2
Mitral incompetence	3
Transposition of the great vessels	20
Corrected transposition of the great vessels	3
Primary pulmonary hypertension	3
"Idiopathic" dilatation of the pulmonary artery	5
Uncertain diagnosis	4
Total	668 (+ 74)

Embryopathy

In 10 of our 668 cases, the mother had contracted a virus infection during the first months of pregnancy: in nine, the infection had been rubeola, and in one, mumps.

Heredity

A certain familial predisposition to congenital heart disease was demonstrated by McKeown *et al.* (455) but could not be shown in the series studied by Richards *et al* (577). In our series, we had two pairs of twins with patent ductus arteriosus, as well as one patient whose sister had earlier been operated on for this malformation. In one case of coarctation of the aorta, another member of the family had the same cardiac anomaly. The series also includes two siblings with tetralogy of Fallot. An additional four patients each had a sibling with congenital heart disease, but of a type different from that of the patient in question

6 Pulmonary Stenosis with Normal Aortic Root

NORMALLY, systolic pressure is the same in the pulmonary artery and in the right ventricle. If systolic pressure is lower in the former than in the latter, pulmonary stenosis is present. Recording of the pressure in the pulmonary artery and in the right ventricle by means of cardiac catheterization is therefore the most reliable means of diagnosing a stenosis. Consequently, it is understandable that pulmonary stenosis with normal aortic root became recognized as a clinical entity and more generally diagnosed only after the introduction of cardiac catheterization.

It is true that single, typical cases had been described earlier (107, 413, 496, 650, 170) In fact, as early as 1817, Meckel (483) gave an account of the pathologic anatomy of this malformation. It was not, however, until 1948-1950 and later that more detailed accounts of the clinical features of the disease were issued from different quarters (2, 8, 132, 206, 299, 408, 472, 539, 592).

The term pulmonary stenosis with normal aortic root is that of Wood (713). Many other terms have been used, among them are pure (299), isolated (472), without over-riding aorta (408), simple (132), uncomplicated (206, 496) and with intact ventricular septum (8, 132).

All degrees of pulmonary stenosis are found. The pressure gradient between the pulmonary artery and the right ventricle may be only around 10 mm Hg, whereas in other cases, a pressure of up to 300 mm Hg has been recorded in the right ventricle, with a normal or slightly decreased pressure in the pulmonary artery As a rule, the lowest pressure gradient indicative of a stenosis has been considered to be 10 mm Hg, but some authors have given 15 mm Hg as the borderline figure (295). In 29 healthy children and eight adults, we found that the pressure gradient did not as a rule exceed 5 mm Hg at rest Only in five individuals did it amount to 6 to 9 mm Hg During exercise, on the contrary, the pressure gradient was larger; in one case it increased from 5 to 21 mm Hg. This can be explained by a shortening of the ejection time of the ventricle, which produces such a large increase in the velocity of flow through the orifice that the pressure of velocity comes into effect, and the pressure recorded in the pulmonary artery is therefore too low (364). Provided that the flow through the pumonary orifice is of ordinary size, a pressure gradient in excess of 10 mm Hg at rest implies that a narrowing must be present at some site between the cavity of the right ventricle and the main trunk of the pulmonary artery. The stenosis may then be in the infundibular part of the ventricle or in the pulmonary orifice. A schematic representation of the different types of pulmonary stenosis is shown in Figure 126.

In valvular stenosis, the cusps are fused

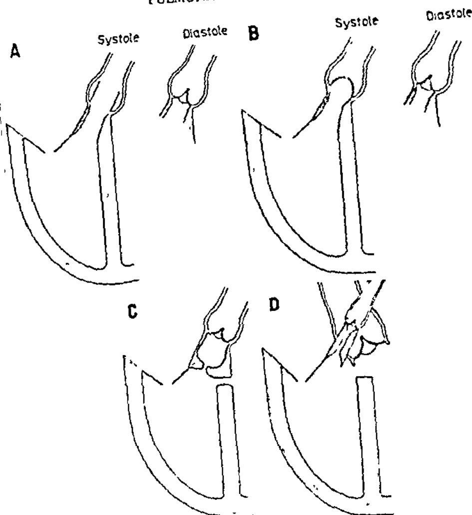


Fig 126 —Schematic representation of different types of pulmonary stenosis A, normal

septal defect

into a membrane with a hole, which is not infrequently eccentrically placed. During systole, the membrane protrudes into the main trunk of the pulmonary artery like a dome, but during diastole it is thrust back toward the ventricle, and the dome becomes flattened or inverted In some cases, the membrane is thin and is considerably displaced during both systole and diastole. In other cases it is thick and almost immovable. Infundibular stenosis is circumscribed and is situated on the borderline between the main part of the right ventricle and the infundibulum The latter forms a cavity, the so-called third ventricle. It is only in combination with an over-riding aorta that the whole infundibulum may be stenosed and form a narrow channel which extends as far up as the valve. A more detailed anatomic description is given on page 191, in connection with the angiocardiographic findings

Pulmonary stenosis with normal aortic root is frequently associated with other cardiac malformations. The combination with an atrial or ventricular septal defect is particularly common and important.? With a large left to right shunt, there is often a pressure gradient between the right ventricle and the pulmonary artery, although there is no malformation of the infundibulum or the valve. The largest pressure gradient we observed was 19 mm Hg, in a case with a very large atrial septal defect. If the gradient had been more than 20 mm Hg, an anatomic stenosis could have been demonstrated by angiocardiography. The great velocity of flow affects the pressure recorded through a catheter with an opening in its tip (364). Higher pressure is recorded in the branches of the pulmonary artery than in its main trunk (see Fig. 116, p. 120). This is because the velocity of flow is greatest just distal to the pulmonary orifice and is greater in the center of the vessel than beside its wall, which explains the fact that different pressures can be measured in various parts of the main trunk Consequently, no pressure gradient is found in many cases with a large left to right shunt A relative stenosis can be conceived to arise by the main trunk of the pulmonary artery being dilated, whereas the valvular ring retains its ordinary width

A ventricular septal defect is not uncommonly combined with stenosis of the infundibulum. The stenosis sometimes is so slight that a left to right shunt is still present through the defect. In severe stenosis, on the contrary, a right to left shunt occurs, and the condition is then difficult to distinguish from tetralogy of Fallot (292). The only distinguishing feature is the over-riding of the aorta. The clinical and hemodynamic conditions may be identical. In many cases, the shunt is so small at rest that the defect cannot be demonstrated by means of blood-gas analysis, but it is usually possible to establish by means of angiocardiography with injection into the right ventricle, owing to the artificial rise in pressure during diastole that is caused by the injection)

In the presence of an atrial septal defect, the direction of the shunt is determined partly by the resistance of the ventricle during diastole. With an uncomplicated defect of this nature, the resistance is lower in the right ventricle than in the left, and the shunt is therefore from left to right (see further, p. 442). With hypertrophy or right ventricular failure, there is increased resistance in the right ventricle and the shunt may then be reversed. An atrial septal defect combined with slight pulmonary stenosis may exhibit a left to right shunt, but when stenosis is pronounced, a right to left shunt occurs. In patent foramen ovale with complete functional closure by the septum, the shunt can take place in one direction only, i.e., from right to left. The shunt may be sufficiently large to cause severe cyanosis. In the French literature, this condition is known as the trilogy of Fallot (326, 620).

Pulmonary stenosis with normal aortic root may be divided into the following types.

1 With intact ventricular and atrial septa
2. With ventricular septal defect and intact atrial septum
   a) With left to right shunt
   b) With right to left shunt
3. With patent foramen ovale and intact ventricular septum
   a) Without shunt
   b) With right to left shunt
4. With atrial septal defect and intact ventricular septum
   a) With left to right shunt
   b) With right to left shunt
5 Combination of ventricular septal defect and atrial septal defect or patent foramen ovale, respectively

In ventricular or atrial septal defect with a left to right shunt combined with pulmonary stenosis (groups 2a and 4a), there is an increase in the pulmonary blood flow. Clinically, these cases belong to the septal defects. In cases of this nature, it is suitable to distinguish between septal defect with pulmonary stenosis, and pulmonary stenosis with septal defect. Obviously, it is difficult to draw a line between these two groups. The decisive factor is which of the two features is of greater importance· the stenosis or the shunt. We have classified

group 2b with a large right to left shunt and cyanosis at rest under tetralogy of Fallot, even when the aortic root is in the normal position Cases with a small right to left shunt, without cyanosis at rest and with arterial oxygen saturation over 85 per cent, have been classified as pulmonary stenosis with ventricular septal defect. This may appear to be an arbitrary classification Cyanosis often appears only when the patient becomes older, a fact which further illustrates the difficulty of classifying certain cases. We are entirely in agreement with Nadas (502) that the term tetralogy of Fallot is unsuitable, but—irrespective of the name given to a pathologic condition—the difficulty of classifying borderline cases remains The congenital anomalies are so heterogeneous—with respect both to anat- [illegible]

appropriate category In our opinion, the foregoing classification is the most feasible from the clinical viewpoint. Examples of the different types of pulmonary stenosis are shown in Figure 127.

Hemodynamically, pulmonary stenosis is characterized by increased resistance to emptying of the right ventricle, which therefore becomes hypertrophied The diastolic filling of a hypertrophied right ventricle requires increased pressure. Consequently, a rise in pressure occurs in the right atrium as well, particularly during auricular systole The atrium becomes hypertrophied When stenosis is only moderate, the stroke volume may remain normal both at rest and on exertion, owing to the ability of the heart to increase the ejection pressure of the right ventricle. In severe stenosis, on the contrary, the stroke volume is small even at rest and decreases still more on exertion The low cardiac output and the high arteriovenous oxygen difference explain the slight peripheral cyanosis that can be observed in these cases. In septal defect with a right to left shunt, the pulmonary blood flow is still further decreased and there is arterial oxygen unsaturation Severe right ventricular hypertrophy may result in a sufficient rise in pressure in the right atrium to cause a right to left shunt. With right ventricular failure, the shunt increases in size As a rule, cardiac decompensation develops only when the patient reaches adult age, but in extremely severe stenosis it may occasionally appear as early as the first year of life (278, 502, 606). More marked cyanosis usually is a late symptom in pulmonary stenosis with atrial septal defect.

In our series, 70 cases could be referred to the pulmonary stenosis group according to the criteria listed. The anatomic diagnosis is recorded in Table 3. In 61 of these cases, the diagnosis was confirmed by angiocardiography with injection into the right ventricle. The age distribution and the physical development are shown in Figure 128. The patients consisted of 37 boys and 33 girls.

CLINICAL FEATURES

Abrahams and Wood (2) have given a detailed description of the signs and symptoms. They criticized earlier publications on this subject (206, 462) and stated that "an accurate clinical diagnosis is nearly always possible at the bedside" It is, in fact, due largely to the contributions of the British cardiologists (Wood (713), Leatham (414), and others) that it is now possible to express complete agreement with [illegible]

[illegible] to discovery of the anomaly. In Sweden, where the majority of children are under continuous health control from birth onward, such cases are detected very early. In our series, the heart disease was diagnosed during the first year of life in two-thirds of the cases and before the age of 7 in the remainder. In practically every instance, the condition was discovered in the course of a routine medical examination.

The children were usually normally developed with respect to weight and height (Fig 128) Clinical symptoms were present in 31 patients, but only four of them were

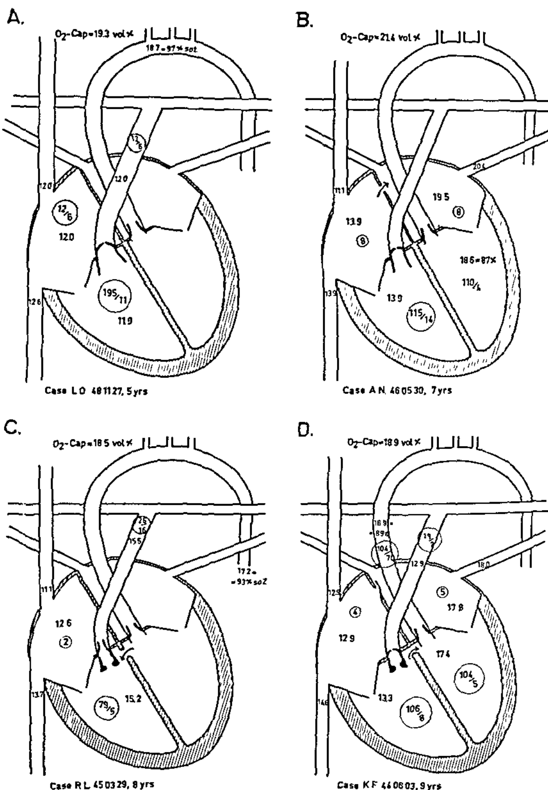


Fig. 127.—Examples of hemodynamics in different types of pulmonary stenosis. The anatomic picture of the stenosis and the ventricular septum was established by selective angiocardiography. *A*, valvular stenosis with no shunt. *B*, valvular stenosis with right to left interatrial shunt. *C*, mild, infundibular stenosis with a third ventricle, combined with ventricular ventricular septal defect *D*, combined valvular e, with right to left shunt through a ventricular (*continued*).

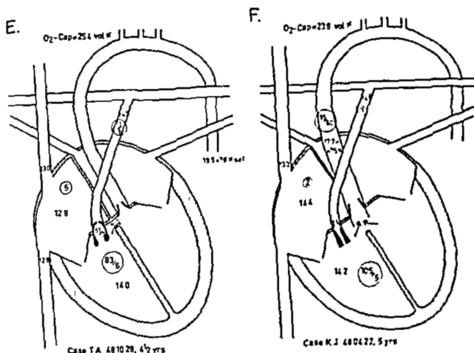


Fig. 127 (*cont.*) —*E*, coincident severe valvular and mild infundibular stenosis with large right to left shunt through a ventricular septal defect. Clinically, the picture was that of a tetralogy of Fallot, so the case was classified under this diagnosis, although angiocardiography showed a normal aortic root *F*, for comparison, a case of tetralogy of Fallot, in which over riding aorta was demonstrated at angiocardiography

TABLE 3 —Anatomic Diagnosis in 70 Cases* of Pulmonary Stenosis with Normal Aortic Root

Anatomy of Septa	Anatomy of Stenosis			Total
	Valvular Stenosis	Infundibular Stenosis	Valvular + Infundibular Stenosis	
Intact septa . . .	39†	4	2	45
Ventricular septal defect . . .	0	5‡	4	9
Atrial septal defect (or patent foramen ovale)	10	1	1	12
Ventricular and atrial septal defect (or patent foramen ovale).	0	2	2	4
Total	49	12	9	70

*An additional 14 adult patients from other clinics were studied with selective angiocardiography only 11 had valvular stenosis and three infundibular stenosis An additional seven patients had atrial septal defect [illegible]

[illegible]

‡ [illegible] one case with atresia of the left main branch of the *pulmonary artery*

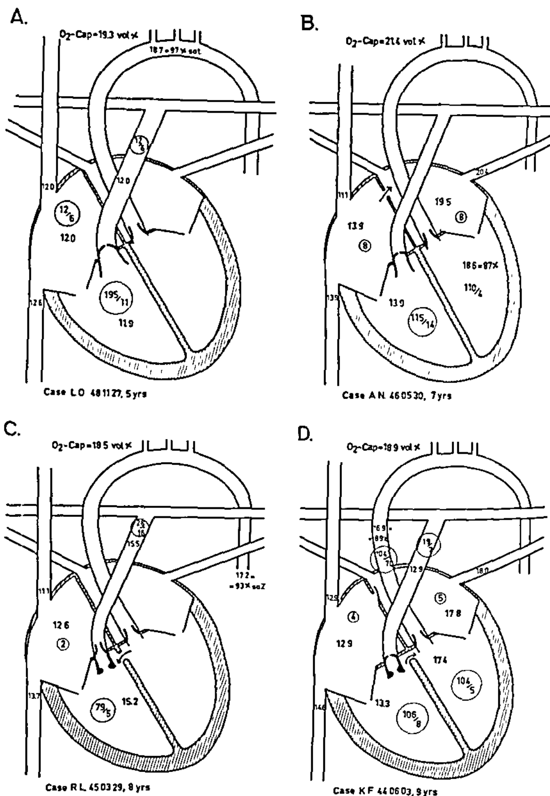


Fig. 127.—Examples of hemodynamics in different types of pulmonary stenosis The anatomic picture of the stenosis and the ventricular septum was established by selective angiocardiography. *A*, valvular stenosis with no shunt. *B*, valvular stenosis with right to left interatrial shunt. C, mild, infundibular stenosis with a third ventricle, combined with ventricular septal defect and left to right shunt, classified as ventricular septal defect *D*, combined valvular and infundibular stenosis of more severe degree, with right to left shunt through a ventricular septal defect—normal position of the aortic root (*continued*)

severely incapacitated The most common symptoms were increased fatigability (23 cases), dyspnea (20 cases), and slight cyanosis on exertion (18 cases). Two of the patients with an interatrial shunt were squatters, both had decreased arterial oxy-

had pulmonary stenosis and intact septa. Since 8 months of age she had increasing hepatomegaly and edema, as well as dyspnea even at rest

GIRL, AGED 9 YEARS (B H 470514) —She had valvular stenosis, atrial septal defect, and a large right to left shunt. She presented the

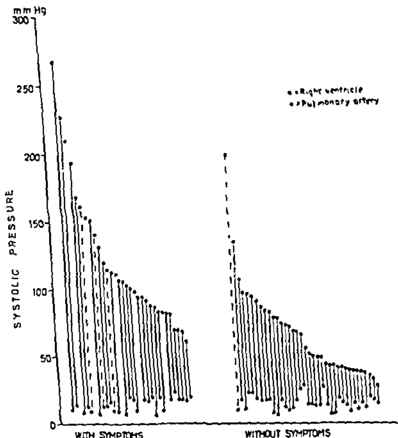


FIG. 129.—Pressure gradient between right ventricle and pulmonary artery. Systolic pressure in ventricle and artery are connected by line. Broken line denotes that pulmonary artery pressure was not measured, PCA pressure was recorded instead Comparison is made between patients with clinical symptoms and those without symptoms Age distribution was same in the two categories

gen saturation, but only one was deeply cyanotic The incidence of cyanosis is higher in adult patients (2, 8, 132). In a series of 10 adults, we observed marked cyanosis in three cases)

The salient features in the four severely incapacitated patients were as follows

GIRL, AGED 18 MONTHS (M P 550510) —She

a typical squatter. The arterial oxygen saturation at rest was 64 per cent.

GIRL, AGED 2 YEARS (S R 531225) —There were valvular pulmonary stenosis and intact septa. Dyspnea and peripheral cyanosis had

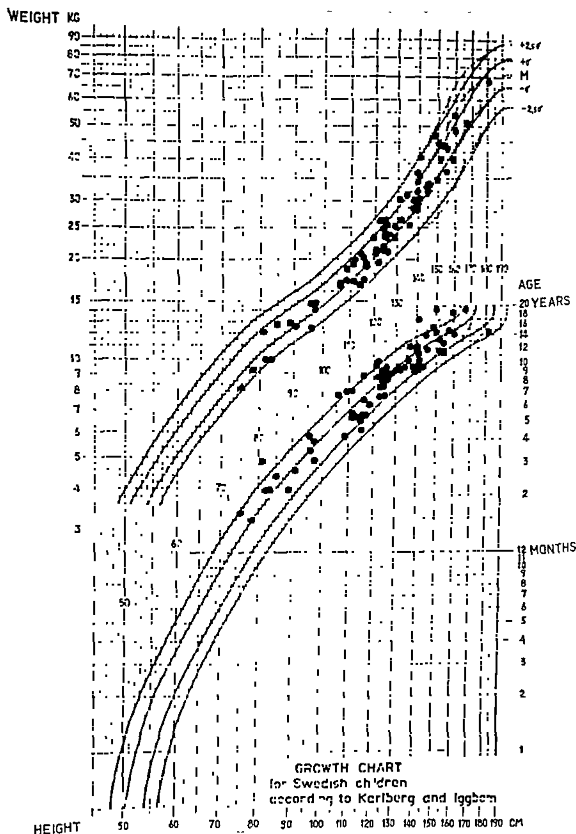


Fig 128.—Correlation of age, height, and weight in pulmonary stenosis Physical development lies within normal limits of variation for Swedish children (Relevant growth chart was placed at our disposal by courtesy of Drs P Karlberg and S Iggbom [374, 375])

severely incapacitated. The most common symptoms were increased fatigability (23 cases), dyspnea (20 cases), and slight cyanosis on exertion (18 cases). Two of the patients with an interatrial shunt were squatters, both had decreased arterial oxy- had pulmonary stenosis and intact septa. Since 8 months of age she had increasing hepatomegaly and edema, as well as dyspnea even at rest.

GIRL, AGED 9 YEARS (B H 470514).—She had valvular stenosis, atrial septal defect, and a large right to left shunt She presented the

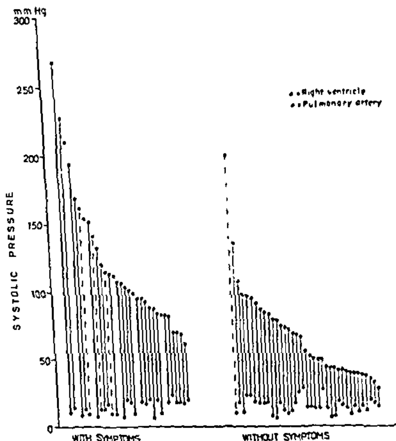


Fig 129.—Pressure gradient between right ventricle and pulmonary artery Systolic pressure in ventricle and artery are connected by line Broken line denotes that pulmonary artery pressure was not measured, PCA pressure was recorded instead Comparison is made between patients with clinical symptoms and those without symptoms Age distribution was same in the two categories

gen saturation, but only one was deeply cyanotic. The incidence of cyanosis is higher in adult patients (2, 8, 132) In a series of 10 adults, we observed marked cyanosis in three cases)

The salient features in the four severely incapacitated patients were as follows.

GIRL, AGED 18 MONTHS (M P 550510) —She same clinical picture as that in severe tetralogy of Fallot Cyanosis, which had appeared during her first year of life, was extremely deep at the present examination She had marked hourglass nails and clubbing of the fingers and was a typical squatter The arterial oxygen saturation at rest was 64 per cent

GIRL, AGED 2 YEARS (S.R 531225) —There were valvular pulmonary stenosis and intac septa Dyspnea and peripheral cyanosis ha

been present since the age of 14 months. At examination, she was found to have dyspnea at rest and enlargement of the liver, but no edema. She spent most of the day in bed.

BOY, AGED 3½ YEARS (O.J. 511509).—The patient had valvular stenosis and a patent foramen ovale with a small right to left shunt, had slight peripheral cyanosis and liver enlargement, but no edema He could manage only two flights of stairs

The relation between the degree of stenosis and the clinical symptoms is apparent from Figure 129 With a systolic pressure below 70 mm Hg, all patients except one were asymptomatic, and with a pressure above 100 mm Hg, all except three had symptoms. Of those with a systolic pressure between 100 and 70 mm Hg, there were as many with symptoms as without them.

PHYSICAL WORKING CAPACITY

Obviously, it is difficult to make an exact evaluation of the symptoms, especially in children. The knowledge of an existing heart disease may induce symptoms, and more or less normal fatigue may be ascribed to the cardiac condition On the other hand, the symptoms may be underestimated Consequently, it is desirable to determine the physical working capacity with objective methods (see p 105), but this is feasible *only in older children and* adults Figure 130 shows the results of such tests in 12 children, as well as in 11 adults who are not otherwise included in our series A positive correlation exists between the degree

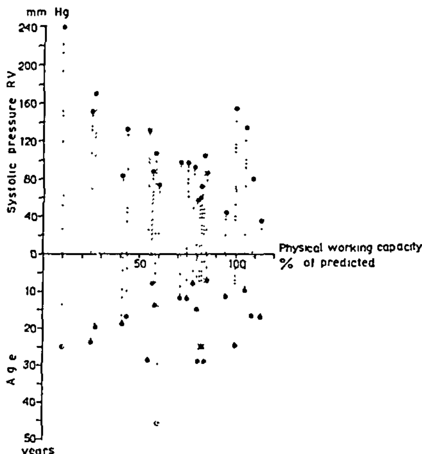


FIG. 130.—Pulmonary stenosis Physical working capacity, per cent of predicted (see p 106), in relation to systolic pressure in right ventricle and age of patient The higher the pressure in the right ventricle, the lower is the working capacity There is also a tendency to decreasing working capacity with rising age The individual variations are, however, large, and patients with severe stenosis may have a normal physical working capacity even at 25 years of age.

of stenosis* and the decrease in physical working capacity, although this may be good even in severe stenosis. The ability of the right ventricle to maintain a normal stroke volume despite the increased resistance in the outflow tract varies appreciably from case to case. Thus, with stenosis of the same degree of severity, one patient may become seriously incapacitated in child-hood whereas another ...

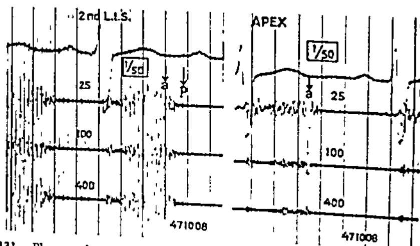


Fig 131.—Phonocardiogram in severe valvular pulmonary stenosis Boy, aged 6 (T.J. 471008) Right ventricular pressure 270/8 mm Hg, pulmonary artery pressure 13/8. The murmur is late systolic, diamond shaped and continues with small amplit... component of the second ... note standa... sound, L I S ...

(615), but cases have been described in which there was relative freedom from symptoms even at 60 years of age despite a systolic pressure in the right ventricle of between 70 and 100 mm Hg (702)

Physical Signs

The *blood pressure and peripheral arterial pulse* were normal except in very severe stenosis with intact septa. In such cases, the low stroke volume results in a small pulse amplitude, and a pathologic venous pulse may be observed

Palpation of the precordium.—The right ventricular hypertrophy which is the most prominent sign of the disease can easily be diagnosed by palpation. The apex beat is displaced backward and is not palpable, or only faintly, whereas the hypertrophied right ventricle strikes, at each heart beat, against the thoracic wall and causes a characteristic parasternal or substernal lift (2). This important sign is present in all except the mildest cases.

Precordial bulge was visible in 11 cases, but in only one of them was it prominent. This is in good agreement with the roentgenologic observations, which usually disclosed a heart of normal size or only slightly enlarged

Sounds and murmurs.—Nothing noteworthy was found in the first heart sound. In three cases, an early systolic sound was heard over the second left interspace (275, 417, 433); on the phonocardiogram it occurred 0.05 ... the first ...

*The degree of severity of stenosis cannot be expressed only by means of the pressure gradient, but is also determined by the stroke volume and ejection time In cardiac incompetence, the stroke volume decreases and the ejection time is prolonged. Consequently, in patients with very low physical working capacity, the stenosis is more marked than can be inferred from the pressure in the right ventricle

pulmonary stenosis, with a pressure gradient between the right ventricle and the pulmonary artery amounting in the respective cases of 12, 26, and 33 mm Hg (see Fig. 132, *D*). The sound was synchronous with the start of upstroke in the pulmonary electrokymogram (p. 168). Like Leatham and Vogelpoel (417), we were unable to detect any early systolic sound in any of the cases of severe pulmonary stenosis, although the poststenotic dilatation was marked. Nor was the pathologic accentuation of the first sound described by Reinhold and Nadas (554) audible or recorded in our cases. In severe stenosis, an atrial sound may be heard (2, 275, 413, 418, 624, 675), but this applied in only one of our patients

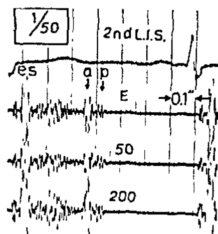


Fig. 132.—Phonocardiogram in mild valvular pulmonary stenosis Girl, aged 9 (C N. 450828). Right ventricular pressure, 42/8 mm Hg, pulmonary artery pressure, 16/8. The murmur is low-frequent, and the greatest intensity is in the first half of systole Note the early systolic sound (*es*) The interval between the aortic and pulmonic components (*a* and *p*) of the second sound is rather short Boxed figure denotes degree of amplification, other figures denote standard frequency of the filters. *L I S*, left interspace

The pulmonary component of the second heart sound exhibited a decreased amplitude in severe cases of both valvular and infundibular pulmonary stenosis

In the most severe cases of valvular stenosis, *the* second heart sound over the pulmonary area cannot be detected with the stethoscope. This is because the aortic component is masked by the systolic murmur and the pulmonary component is so faint that it cannot be recognized. In most cases, both components can be recorded on the phonocardiogram.

Figure 131 is a phonocardiogram in a case of valvular pulmonary stenosis; it was recorded with the microphone over the second left interspace. The position of the aortic component was determined by phonocardiograms over the apex and the aortic area; it occurred before the end of the systolic murmur, which was then characterized by maximum amplitudes. *In all our cases, the pulmonary component was delayed and appeared 0.06–0 10 sec after the beginning of the aortic component. In cases with a moderate or marked pressure gradient, its amplitude was only one-fourth to one-fifth of that of the aortic component* In the mild cases with a right ventricular pressure below 70 mm Hg, *both components of the second heart sound can be heard and seen without difficulty in the phonocardiogram. The interval between the components is shorter and their amplitude is approximately the same* (Fig 132).

In infundibular stenosis, the degree of splitting of the second sound is the same as in valvular stenosis. Since in these cases the systolic murmur ends with the aortic component, the splitting is often easier to hear (see following paragraphs). On auscultation at the left sternal border over the third or fourth interspace, the pulmonary component is audible as a faint extra sound after the end of the murmur. Figure 133 shows three cases of infundibular stenosis in which both the aortic and the pulmonary components of the second sound were recorded.

The systolic murmur is one of the most important clinical signs of the disease In all cases of valvular stenosis, it has a *definite maximum over the second* left interspace, it is protracted, harsh and of high frequency and intensity, grade 4–6

Phonocardiographically, the systolic murmur in valvular stenosis has the following characteristics (Figs. 131 and 132) With

...ere stenosis, the murmur is diamond-...aped (416) with the maximal intensity ...te in systole In 27 of our 31 cases with a ...ght ventricular systolic pressure of 70–230 ...m Hg, the highest amplitude of the sys-...lic murmur was recorded in the latter half ...f the interval between the first heart sound ...nd the pulmonary component of the sec-...nd sound

In mild stenosis, the intensity of the ...murmur is greatest in the first half of

the intensity of the murmur. The decisive factors are the velocity and size of the flow through the pulmonary orifice and the distance between the blood jet and the thoracic wall. In three of the severest cases the murmur was very faint, grade 2-3. Two of these patients (M.P. 550510 and S.R. 531225) had right ventricular failure. In this condition, the stroke volume is decreased and the duration of systole prolonged, with a resulting decrease in the velocity of flow through

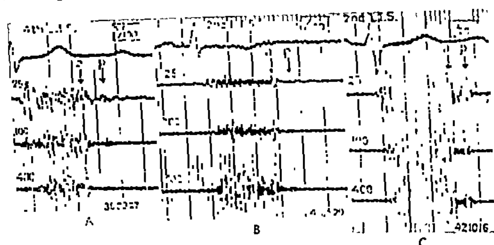


Fig. 133 —Phonocardiogram in three cases of infundibular stenosis. A, boy, aged 15 (I B [illegible]

components of the 2nd sound, L I S, left interspace

systole (Fig 132) This applied in 17 of our 19 cases with a systolic pressure in the right ventricle below 70 mm Hg Except in the mildest cases, the murmur continued beyond the aortic component of the second sound This is often masked by the murmur, and can therefore be observed only on the phonocardiogram recorded over the apex region

The more severe the stenosis, the louder the murmur But it is not the degree of severity of stenosis alone which determines

the stenosis. In the third case (B.H. 470514), there was an extremely large right to left shunt at the atrial level and, consequently, a very small pulmonary flow. In addition, the thoracic wall was deformed, and the blood jet through the pulmonary orifice was directed backward Plentiful lung tissue lay between the pulmonary artery and the anterior thoracic wall (Fig 134, B). The murmur was only grade 3, despite systolic pressure of 210 mm Hg in the right ventricle. In such cases the condi-

pulmonary stenosis, with a pressure gradient between the right ventricle and the pulmonary artery amounting in the respective cases of 12, 26, and 33 mm Hg (see Fig. 132, *D*). The sound was synchronous with the start of upstroke in the pulmonary electrokymogram (p. 168). Like Leatham and Vogelpoel (417), we were unable to detect any early systolic sound in any of the cases of severe pulmonary stenosis, although the poststenotic dilatation was marked. Nor was the pathologic accentuation of the first sound described by Reinhold and Nadas (554) audible or recorded in our cases. In severe stenosis, an atrial sound may be heard (2, 275, 413, 418, 624, 675), but this applied in only one of our patients

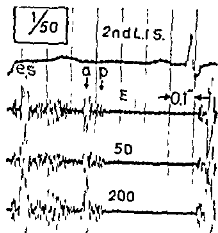


Fig. 132.—Phonocardiogram in mild valvular pulmonary stenosis Girl, aged 9 (C N. 450828) Right ventricular pressure, 42/8 mm Hg, pulmonary artery pressure, 16/8 The murmur is low-frequent, and the greatest intensity is in the first half of systole Note the early systolic sound (*es*) The interval between the aortic and pulmonic components (*a* and *p*) of the second sound is rather short Boxed figure denotes degree of amplification, other figures denote standard frequency of the filters, *L I S*, left interspace

The pulmonary component of the second heart sound exhibited a decreased amplitude in severe cases of both valvular and infundibular pulmonary stenosis

In the most severe cases of valvular stenosis, the second heart sound over the pulmonary area cannot be detected with the stethoscope. This is because the aortic component is masked by the systolic murmur and the pulmonary component is so faint that it cannot be recognized. In most cases, both components can be recorded on the phonocardiogram.

Figure 131 is a phonocardiogram in a case of valvular pulmonary stenosis, it was recorded with the microphone over the second left interspace. The position of the aortic component was determined by phonocardiograms over the apex and the aortic area; it occurred before the end of the systolic murmur, which was then characterized by maximum amplitudes In all our cases, the pulmonary component was delayed and appeared 0.06–0.10 sec after the beginning of the aortic component. In cases with a moderate or marked pressure gradient, its amplitude was only one-fourth to one-fifth of that of the aortic component. In the mild cases with a right ventricular pressure below 70 mm Hg, both components of the second heart sound can be heard and seen without difficulty in the phonocardiogram. The interval between the components is shorter and their amplitude is approximately the same (Fig. 132).

In infundibular stenosis, the degree of splitting of the second sound is the same as in valvular stenosis. Since in these cases the systolic murmur ends with the aortic component, the splitting is often easier to hear (see following paragraphs). On auscultation at the left sternal border over the third or fourth interspace, the pulmonary component is audible as a faint extra sound after the end of the murmur Figure 133 shows three cases of infundibular stenosis in which both the aortic and the pulmonary components of the second sound were recorded

The systolic murmur is one of the most important clinical signs of the disease In all cases of valvular stenosis, it has a definite maximum over the second left interspace, it is protracted, harsh and of high frequency and intensity, grade 4–6.

Phonocardiographically, the systolic murmur in valvular stenosis has the following characteristics (Figs 131 and 132) With

dibular stenosis showed that the systolic murmur started immediately after the first heart sound and, in most cases, ended abruptly with the aortic component of the second sound[1] (Fig. 133). This was succeeded by a pause varying from 0.05 to 0.10 sec—i.e., of the same duration as in valvular stenosis—followed by the faint pulmonary component of the second heart sound. In patients with a ventricular septal defect—particularly with a large left to right shunt, these cases being classified as ventricular septal defects associated with pulmonary artery stenosis—the systolic murmur was often extremely loud. In a few such cases (Fig. 133, C) the systolic murmur was among the loudest that can be heard. In these cases, very slight variations were found in the interval between the two components of the second heart sound.

It can be shown, by simultaneous phonocardiography and recording of the intracardiac pressure curves, that the weak, delayed pulmonary component is caused, in both valvular and infundibular stenosis, by closure of the pulmonary valve. It coincides in time with the notch in the pulmonary artery curve. These observations will be discussed in more detail on page 185 (Fig. 179).

We have seen one case of supravalvular pulmonary stenosis due to a membrane about 1.5 cm above the valve. Figure 135 shows the phonocardiogram in this case. An extra sound which, in view of the appearance of the pressure curve occurred too late to be the pulmonary component of the second heart sound, was recorded consistently 0.09 sec after the second sound. This extra sound may have been caused by the membrane in question (see Fig. 213, p. 221)!

In 1954, Mannheimer and Jonsson (471) put forward the following explanation of the observations made.

The pulmonary valvular stenosis causes a prolongation of right ventricular systole (Fig. 180), due to the great resistance to ejection of the stroke volume. This explains the wide splitting of the second sound and the long systolic murmur which continues after the aortic component. The maximal intensity of the murmur is in late systole. Only in very mild stenosis is the murmur early systolic and ends before the aortic component. The stenosed valve is like a membrane, inverted toward the infundibulum during diastole. In the beginning of systole, it protrudes into the pulmonary artery like a dome. This movement involves the displacement of a fairly large quantity of blood without the passage of any blood through the stenosis. This flow and, consequently, the systolic murmur must be somewhat delayed.

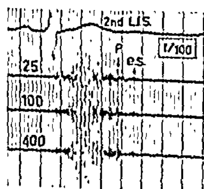


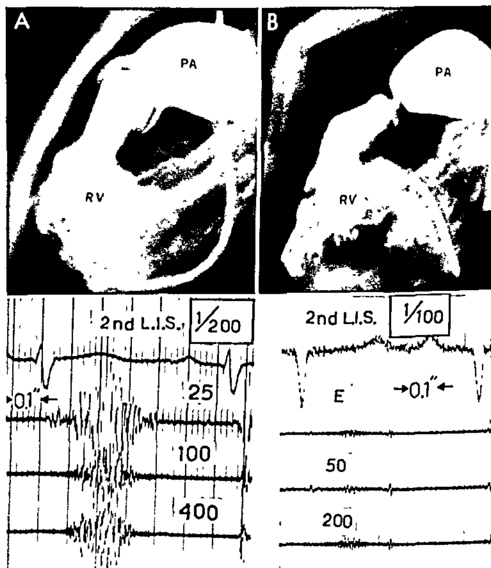
Fig. 135.—Phonocardiogram in pulmonary stenosis due to a supravalvular membrane. Boy, aged 9 (L.W. 440827) Royal ...

The amplitude of the second pulmonary component is due partly to the pulmonary artery pressure and partly to the anatomy of the valves and their distance from the thoracic wall. Since the valvular membrane is usually only moderately thickened and has a wide range of movement, it is not surprising that a distinct pulmonary component can be observed on the phonocardiogram even in severe stenosis.

It is generally accepted that the infundibulum of the right ventricle is the last part of the heart to be excited and contracted. In normal individuals, both

tion cannot be distinguished from tetralogy of Fallot on the basis of the physical signs (128).

By way of contrast, Figure 134, *A* shows a case of mild pulmonary stenosis (systolic pressure in right ventricle 52 mm Hg) with a grade 6 systolic murmur. The jet was directed forward, and little or no lung tissue lay between the pulmonary artery and the thin thoracic wall. In this case, the murmur was of low frequency, whereas in the former case it was of high frequency. It is possible that a more definite conception of the degree of stenosis would be provided by an improved frequency analysis (456)



...d 10 (B H 470514), with extremely ... left interatrial shunt The decreased flow to the anterior thoracic wall may explain

✓In infundibular stenosis, the systolic murmur is heard lower down at the sternal border, most distinctly in the third and fourth left interspaces

✓Phonocardiograms in the cases of infun

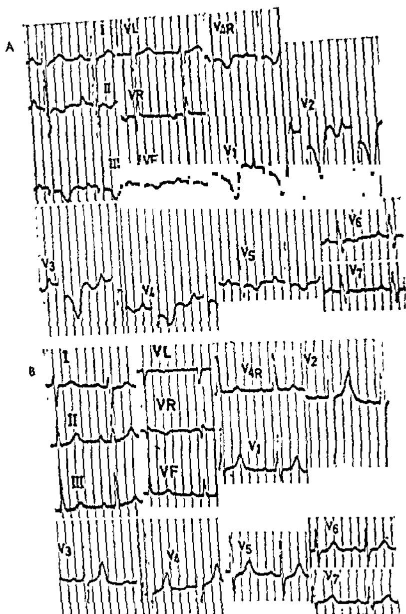


Fig. 136 —Electrocardiogram in: *A*, severe pulmonary stenosis with systolic pressure in right ventricle 270 mm Hg, B, moderate infundibular stenosis with systolic pressure in right ventricle 85 mm Hg

ventricles are emptied of blood almost simultaneously. In infundibular stenosis, on the contrary, the infundibulum—which is converted into a third ventricle—terminates its systole still later than the left ventricle. The murmur occurs in early systole, when blood passes from the lower part of the right ventricle through the stenosis into the third ventricle. When this third ventricle empties its blood through the normal pulmonary valves, no murmur is audible.

It is often possible by simple auscultation to distinguish between valvular and infundibular pulmonary stenosis. By means of phonocardiography, we are now able to make this differentiation with greater certainty.

Electrocardiography

In few other heart diseases are such marked changes indicative of right ventricular hypertrophy found in the ECG as in severe pulmonary stenosis. Figure 136 shows the ECG in severe and in moderate stenosis On the other hand, the ECG may be normal in the mildest cases This applied in 12 of our cases with a systolic pressure in the right ventricle of 35–88 mm Hg. The normal values for the various age groups were determined according to Ziegler (724) The most characteristic change is a tall R wave in V_1 (519) Figure 137 shows, however, that the correlation between the pressure in the right ventricle and the amplitude of the R wave in V_1 is not particularly strong

A negative T wave in V_1–V_4 has been regarded as highly characteristic of severe pulmonary stenosis and often decisive as an indication for operation (132, 326, 478, 527). The correlation between the negative T wave in different precordial leads and the pressure in the right ventricle may be inferred from Figure 137. In children, this correlation is not so evident, since a negative T wave may occur normally in V_4 up to 5 years of age and in V_3 up to 10 years Even if the age factor is taken into account, a negative T wave in the precordial leads is not a reliable indication of the degree of severity of the stenosis. In earlier investigations (132, 361), it has chiefly been patients with severe cyanosis who have exhibited these pathologic T waves.

The hypertrophy and dilatation of the right atrium which occur in severe pulmonary stenosis are reflected electrocardiographically in a tall P wave over the right precordium. It is, however, apparent from Figure 137 that the P wave may be normal even in severe stenosis, whereas peaked P waves may be present in moderate stenosis

Complete right bundle-branch block was observed in one patient, and incomplete right bundle-branch block in eight. The coincidence of incomplete right bundle-branch block and patent foramen ovale in pulmonary stenosis has been pointed out by Selzer *et al.* (592). In only one of our eight cases of incomplete right bundle-branch block could a patent foramen ovale be demonstrated. The low incidence of right bundle-branch block in pulmonary stenosis is in contrast to its frequent occurrence in atrial septal defect with a left to right shunt and dilatation of the right ventricle

Campbell (132) found a prolongation of the P-R interval as a late symptom in severe stenosis Only one of our patients in heart failure had a P-R interval at the normal upper limit.

Thus, in pulmonary stenosis the electrocardiographic changes vary to some extent with the degree of severity of stenosis The individual variations are, however, so large that it is not possible in a particular case to draw any definite conclusions regarding the degree of severity from the electrocardiogram (418) We have observed cases of pulmonary stenosis and intact ventricular septum in which the electrocardiogram was normal despite a right ventricular pressure of 88 mm Hg. In another case, in which there was a right ventricular pressure of 142 mm Hg, there were only inconspicuous electrocardiographic changes, with R in V_1 equaling 1 6 millivolt, P in V_1 equaling 1 millivolt, a normal T wave and S-T interval, and an R S quotient in V_1 equaling 1.3.

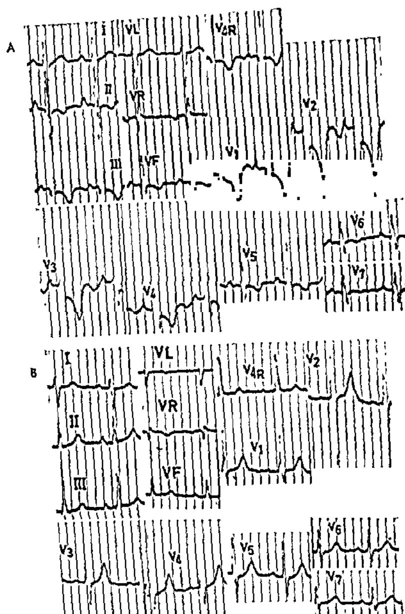


Fig 136.—Electrocardiogram in *A*, severe pulmonary stenosis with systolic pressure in right ventricle 270 mm Hg. *B*, moderate infundibular stenosis with systolic pressure in right ventricle 85 mm Hg

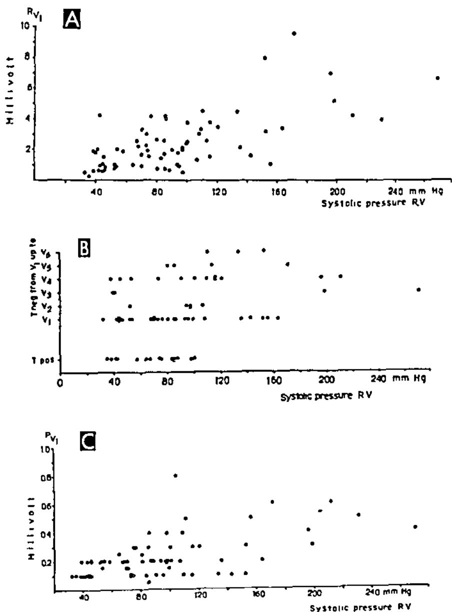


Fig. 137.—*A*, correlation of R wave in V_1 with systolic pressure in right ventricle. *B*, T wave [illegible] Pathologic T wave is not invariably [illegible] exhibited marked cya- [illegible] Filled circles denote cases with intact ventricular septum, and open circles, cases with associated ventricular septal defect.

ROENTGENOLOGIC EXAMINATION

Valvular Stenosis

Detailed knowledge of the roentgenologic findings in valvular pulmonary stenosis with intact ventricular septum is based on experience which dates from slightly less than 20 years ago. The roentgenologic appearance has been described by Laubry et al (411), Dow et al. (206), Greene et al. (299), Healey et al (321), Blount et al. (75), and Saltzman (575), among others.

The poststenotic dilatation of the main trunk of the pulmonary artery is a classic finding (Fig. 138); it was lacking in only two of our patients over 4 years of age (Fig. 140). In infants and small children, such dilatation is probably uncommon. Only two of our 10 patients in these categories had any marked dilatation of the main trunk. Deficient dilatation of this vessel occurred in various degrees of stenosis and could be confirmed by angiocardiography in six cases examined Judging by the dilatation of the central vessels in the hilum or by

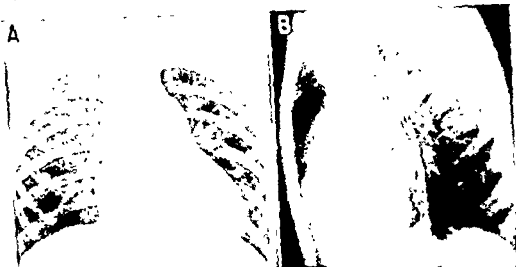


Fig. 138.—Valvular pulmonary stenosis Boy, aged 6 (L O. 481127); see Figure 197 (p. 205). Poststenotic dilatation of main trunk of pulmonary artery, enlargement of right ventricle, and lengthy contiguity of surface to anterior wall of thorax, but no bulge, slight reduction in peripheral vascularity

Our findings, which are based on 62 cases, are in all essentials in agreement with these earlier observations. We found the following features to be characteristic, although none of them is present consistently.

. The main trunk of the pulmonary artery is dilated.

· The peripheral vessels of the lungs are narrow.

. The shape of the right ventricle is hypertrophic and it may be enlarged.

In many cases, there are also enlargement of the right atrium and a prominent auricular appendage

the prominence of the left main branch against the lung parenchyma (Fig. 141), the main branches of the artery were also dilated in 13 cases. In one of them there was dilatation of the main branches, but not of the main trunk. Angiocardiography showed, in addition to valvular stenosis, a definite constriction at the origin of the branches (Fig 139). In all the remaining cases, the central vessels were of normal width or narrow. In our opinion, the pulsations of the main trunk of the pulmonary artery are as a rule considerably larger than those seen in normal cases. This is most evident in the superior and anterior seg-

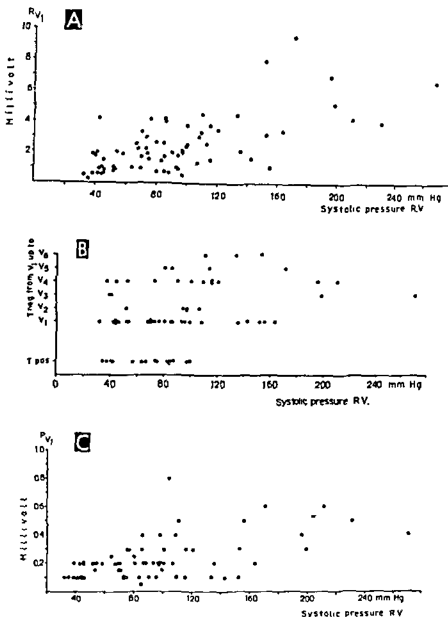


Fig. 137.—*A*, correlation of R wave in V_1 with systolic pressure in right ventricle *B*, T wave in precordial leads in varying degrees of pulmonary stenosis Pathologic T wave is not invariably present in severe pulmonary stenosis All patients were children, and none exhibited marked cyanosis *C*, height of the P wave in varying degrees of pulmonary stenosis Filled circles denote cases with intact ventricular septum, and open circles, cases with associated ventricular septal defect.



Fig 140.—Valvular pulmonary stenosis Boy, aged 8 (L H 450919), see Figure 169 (p 197). No poststenotic dilatation of pulmonary artery, enlargement of right ventricle; right atrium normal, reduction in peripheral vascularity

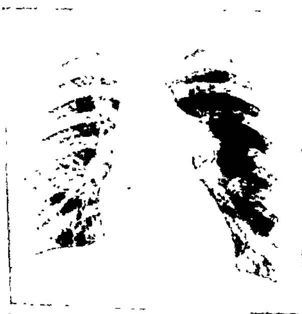


Fig. 141.—Valvular pulmonary stenosis Woman, aged 43 (A F. 100112). Definite dilatation of main trunk and left main branch of pulmonary artery, reduction of vascularity in lungs, slight enlargement of right atrium



Fig 139.—Valvular pulmonary stenosis. Girl, aged 16 (M E. 380224). Fairly large orifice The jet of contrast medium flows centrally, producing a recess-like bulge (*R*) in the main trunk, directly in front of the pericardial reflection (arrow in C). Also considerable stenosis at origin of both main branches (arrows in *B*), with poststenotic dilatation on either side. No increase in width of main trunk.

nary artery can be demonstrated only by angiocardiography.

Peripheral to the hilum, the vascular markings were as a rule less conspicuous than usual. In some cases the vessels were extremely narrow (Fig 142). In one case of severe valvular stenosis, the exceedingly narrow peripheral vessels had an almost reticular arrangement, reminiscent of that seen in collateral circulation (Fig 144). In others, there was only an indeterminate re-

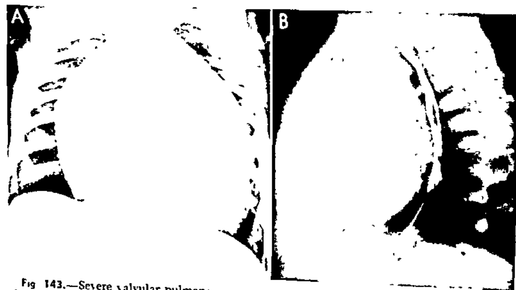


Fig 143.—Severe valvular pulmonary stenosis Girl aged [illegible]
enlargement [illegible]

[illegible]
sis verified at opera[illegible]
demonstration of t[illegible]

duction in the caliber of the vessels, and, particularly in mild stenosis the vascular [illegible]

[illegible] markings and of their normally wide range of variation, a definite statement is not always permissible in such mild cases (e g, Fig 145)

In our experience, the hypertrophy of the right ventricle is best judged in the true lateral projection, it can then be identified by an increased, sometimes bulging contiguity of the heart surface to the anterior wall of the thorax (Figs. 140, 148, 151). The increased curvature of the anterior border of the heart, which is often characteristic in other types of congenital defects with right ventricular hypertrophy, is seldom so distinct in valvular pulmonary stenosis. It may be lacking even in severe stenosis with extremely high ventricular pressure (Fig. 138). Identification of the anterior border of the ventricle may also be made more difficult by the fact that, in the superior part, the right auricular appendage occupies the space between the thoracic wall and the ventricle (Fig. 146).

A distinctly upturned apex is not common in this disease and was observed in only eight of our cases. In 10 other cases there was a suggested curvature of the apex (Fig 147). In the remainder (43 patients) the findings with respect to the shape of the apex were indecisive.

In mild cases of stenosis, the right atrium and its appendage were of ordinary shape and size (Figs 140 and 145). In many of the moderately severe stenoses, and par-

ments of the vessel. The degree of stenosis does not, however, have any marked effect on the size of the pulsations. In electrokymographic tracings, typical changes appear in the shape of the pulsations and are helpful for the diagnosis (p. 168). The pulsations in the vessels at the hilum were small or could not be detected on fluoroscopy Associated movements due to the influence of the main artery were, however, sometimes observed to be present in the left hilar region. In a few cases with dilatation of the branches in the hilum, the pulsations were increased.

The size of the main trunk of the pulmonary artery, judged by the fluoroscopic appearance and the roentgenograms taken in different planes, was in good correlation to the angiographic findings. Our series had only a few cases with such great dilatation of the right ventricle that the greater part of the pulmonary artery was overlapped in standard projections (Fig. 143). Under such conditions, dilatation of the pulmo-



Fig. 142.—Valvular pulmonary stenosis Girl, aged 10 (E I 440429) Marked poststenotic dilatation of main trunk of pulmonary artery, bulging right ventricle, but no increase in contiguity of surface to anterior thoracic wall, very slight dilatation of central vessels and marked decrease in peripheral vascularity, no enlargement of right atrium



Fig 145 —Valvular pulmonary stenosis. Woman, aged 20 (T.Z. 340405). Shape of right ve tricle hypertrophic, with curved anterior surface, considerable poststenotic dilatation of ma trunk of pulmonary artery, no atrial enlargement, vascularity normal



Fig. 146 —Valvular pulmonary stenosis Girl, aged 2 (M S 510423) ...
ment of both ...
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Fig. 144.—Severe valvular pulmonary stenosis and atrial septal defect with large right to left interatrial shunt Girl, aged 10 (B H 470514), cf Figure 195 Considerable hypertrophy of right ventricle, marked dilatation of right atrium, and reduction in vascularity of lungs Owing to extreme stenosis and the reduced flow, poststenotic dilatation of the pulmonary artery is too slight to be apparent The vascular picture partly resembles that in tetralogy of Fallot Deformity of sternum, due to premature synostosis

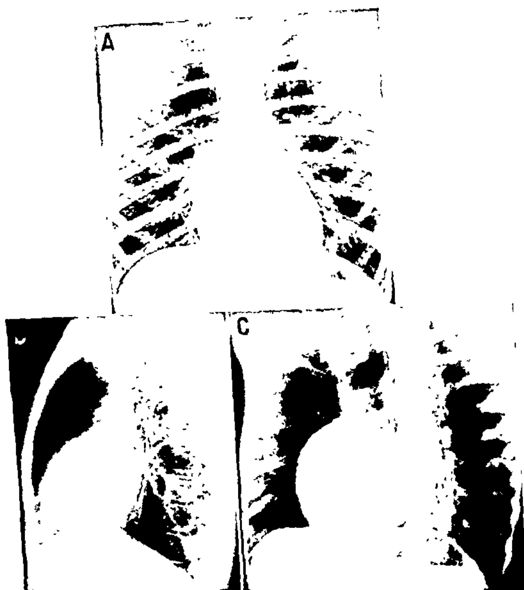


Fig. 148 —Valvular pulmonary stenosis. Girl, aged 9 (B.L. 440127). Moderate poststenotic dilatation of main trunk of pulmonary artery, enlargement of right ventricle and increased contiguity of surface to anterior thoracic wall, enlargement of right atrium, reduction in peripheral vascularity

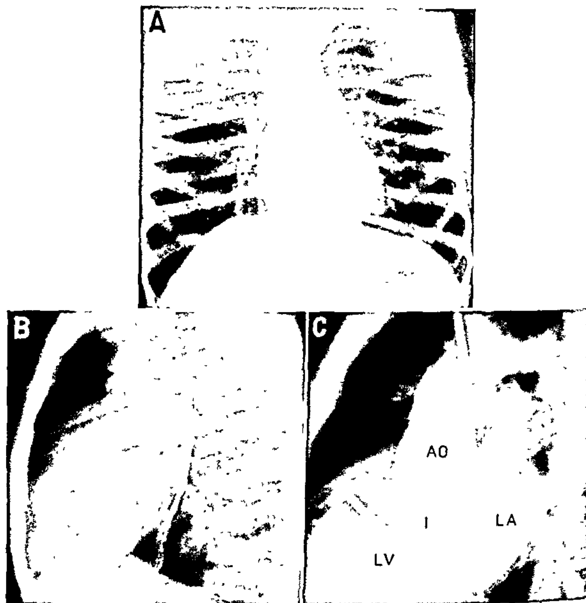


Fig. 147.—Valvular pulmonary stenosis. Boy, aged 9 (J K 441005). *A* and *B*, slight p(stenotic dilatation of main trunk of pulmonary artery, enlargement of right ventricle w large part of surface contiguous to anterior wall of thorax, considerable reduction in periphe vascularity, slight backward bulging of left atrium, probably caused by displacement from enlarged right ventricle. *C*, contrast-filled left atrium is of normal size and displaced dorsa Picture taken in late systole at end of the T wave. Normal residual blood volume in left ven cle, aorta and infundibulum of normal width. *AO*, aorta, *I*, infundibulum, *LA*, left atrium, left ventricle.

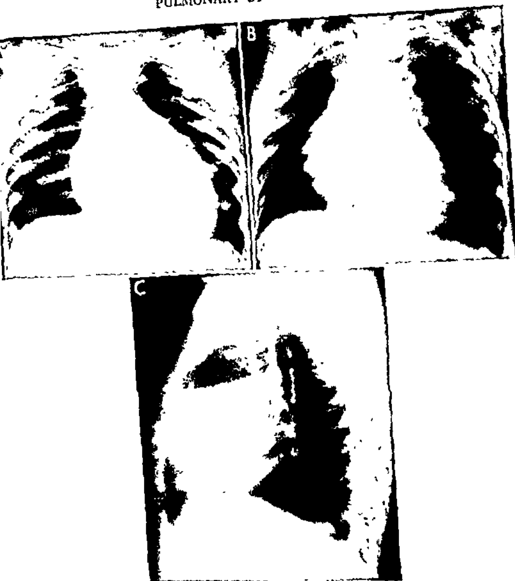


Fig 150.—Valvular pulmonary stenosis. Girl, aged 2 (S R 531225), cf Figure 211. Gross hypertrophy and dilatation of right ventricle and considerable enlargement of right atrium Vascularity of lungs so greatly reduced that the vessel branches are barely identifiable. No demonstrable poststenotic dilatation of pulmonary artery.

ventricular septal defect. From the anatomic point of view, this combination of anomalies includes a number of conditions in which all degrees of stenosis and sizes of septal defect are represented, so that the transition between them is indefinite Hemodynamically, there is the same wide range This matter has been discussed in detail on page 138 and is also dealt with in the section on angiocardiography later in this chapter. Infundibular stenosis without ventricular septal defect has been regarded as rare, but its incidence is presumably higher than was believed earlier.

Infundibular stenosis with or without a ventricular septal defect, and with no left to right shunt, was demonstrated in 17 cases, in five of them there was coincident

ticularly in the severe cases, the atrium was definitely enlarged; the appendage appeared prominent in relation to the adjacent parts, indicating hypertrophy (Fig. 146). In the left oblique projection, marked curvature of the anterior surface of the atrium was visible in several of the severe stenoses (Fig. 148). In severe or moderately severe stenosis, increased activity of the atrium, with deep contractions in presystole, was observed as an expression of increased filling pressure. Although these findings are distinctly visible on fluoroscopy, they are easiest to analyze on the electrokymogram (pp. 175-176)

As stated earlier, a right to left interatrial shunt was present in 12 cases. This produced no essential differences in the roentgenogram. Neither the left atrium nor the left ventricle was enlarged, nor did the width of the aorta differ from the average.

Inappreciable backward protrusion of the left atrium was observed in several cases, it caused a shallow bulge in the esophagus, simulating slight atrial enlargement (Fig. 147) The angiocardiogram in the cases in which this feature was present showed the atrium to be of normal size The phenomenon is presumably to be ascribed to displacement of the atrium or backward tilting of the heart, caused by the right ventricular hypertrophy.

A pathologic increase in heart volume was found in about half of the cases With few exceptions, the increase was moderate and was due to enlargement of the right atrium and ventricle. Gross cardiac enlargement as a sign of heart failure was present in two children, aged 18 months and 3 years, respectively, both with severe stenosis (Figs 143 and 150) In one of these cases the vascularity of the lungs was maximally reduced.



Fig. 149.—Valvular pulmonary stenosis Man, aged 20 (G S 230924). Great enlargement of right ventricle and atrium, very slight poststenotic dilatation of main trunk of pulmonary artery, considerably reduced vascularity in lungs

Supravalvular Stenosis

Our series included one case of supra valvular stenosis (Fig. 151). The roentgenologic findings were identical to those in valvular pulmonary stenosis

Infundibular Stenosis

As already pointed out, infundibular stenosis may be present with intact ventricular septum but is usually accompanied by



Fig. 152.—Infundibular stenosis Girl, aged 12 (G C 410529) As shown in the angiocardiogram (Fig 221, p 231), third ventricle overlaps inferior part of main trunk of the pulmonary artery in frontal projection This was not seen clearly on roentgenologic examination Angulation at level of the pulmonary artery (arrow) is presumably caused by this anomaly.



Fig. 153 (*left*) —Infundibular stenosis Man, aged 30 (K O. 240620). Arrow points to dilated third ventricle, its lower border corresponds to the stenosis, as seen in Figure 220 (p. 230), enlargement of right atrium and ventricle, reduction of vascularity in lungs.

Fig. 154 (*right*) —Infundibular stenosis Boy aged 14 (J R. 280297) [illegible]

227) No [illegible]

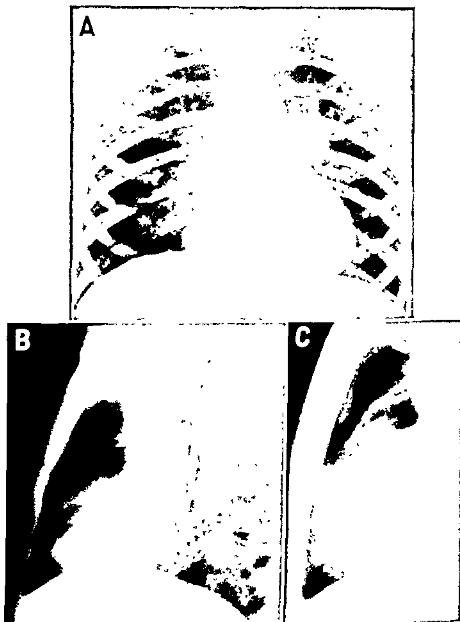


Fig. 151.—Supravalvular pulmonary stenosis Boy, aged 9 (L W 440827), see Figure 213 (p 221). Right ventricle enlarged, with marked curvature of anterior surface (C), very slight post-stenotic dilatation of main trunk of pulmonary artery, reduction in peripheral vascularity.

valvular stenosis. Our observations showed that the roentgenologic appearance is seldom characteristic and that the findings are remarkably variable.

Poststenotic dilatation and conspicuous pulsations in the pulmonary artery combined with decreased vascularity of the lungs—which is often found in valvular stenosis—occurred in two cases only In the others, the pulmonary artery exhibited a normal appearance or was less prominent than usual In addition to the aforementioned, reduced vascularity was observed in four cases, all associated with valvular stenosis. Thus, neither poststenotic dilatation of the pulmonary artery nor changes in the peripheral vessels were nearly as common as in valvular stenosis alone

Evaluation of the main trunk of the pulmonary artery is sometimes difficult because of the partially overlapping third ventricle. A comparison between the roent-



Fig. 156.—Infundibular stenosis. Boy, aged 17 (B H 390705). Slight poststenotic dilatation (arrow in *B*) of main trunk of pulmonary artery. Vascularity of lungs greatly reduced. Right-sided aortic arch Moderate dilatation of right atrium

third ventricle was visible as a moderate local dilatation of the infundibulum. In a third case the infundibular stenosis was seen as a small indentation in the border at the lower part of the infundibulum (Fig. [illegible]

ulum and the pulmonary artery (p. 176).

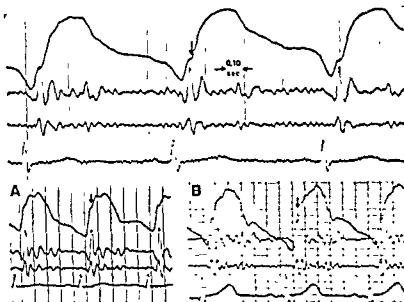
As in valvular stenosis, there is usually slight enlargement of the right ventricle, with an increase in contiguity to the anterior wall of the thorax. In one case, evaluation of the size of the ventricle was prevented by marked rotation of the heart.

Enlargement of the right atrium and its appendage was visible in six cases. In one of them, the right atrium was divided by a



Fig. 155.—Inf ... ...ement of right atrium and ... dilatation clearly ... esophagus.

genogram (Fig. 152) and the angiocardiogram (Fig. 221, p 231) shows that, in the case illustrated, the basal part of the "pulmonary arc" was formed by the superior part of the third ventricle. This fact was not clearly evident on the roentgenographic examination Similar observations were made in several other cases. In only one of them was the third ventricle so large that it was distinctly visualized against both the pulmonary artery and the sinus region of the ventricle (Fig 153). This feature is specific and indicates the presence of infundibular stenosis. In one additional case, a



Figs 158-159 —Pulmonary artery electrokymograms in valvular pulmonary stenosis.

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partition into two partly separate chambers, the cor triatriatum (Fig. 27). There was coincident right aortic arch, and the left subclavian artery caused an indentation in the esophagus (Fig. 155). In this group, as in infundibular stenosis and ventricular septal defect with right to left shunt, a right-sided aortic arch was not uncommon and was found in four additional cases (Fig. 156). Cardiac catheterization gave no grounds for the presence of an intracardiac right to left shunt.

A pathologic increase in heart volume was present in six cases.

In two patients with extremely mild and moderate infundibular stenosis, respectively, the roentgenologic appearance was normal in every respect.

ELECTROKYMOGRAPHY

Valvular Stenosis

On fluoroscopy, large pulsations in the main trunk of the pulmonary artery are a typical although not consistent feature of valvular pulmonary stenosis An analysis of the pattern of the pulsations by electrokymography has shown that characteristic changes in it can be recorded in many cases (15, 397). One of us (571) analyzed these conditions in 53 cases of valvular stenosis with a systolic pressure gradient across the valve ranging from 10 to 260 mm Hg In the series in question, no associated malformation of the heart was present except in five cases, in which there was an atrial septal defect through which a left to right shunt took place. Of the 53 patients, 21 had mild stenosis (pressure gradient across the pulmonary valve less than 50 mm Hg), 18 moderate stenosis (gradient less than 100 mm Hg), and 14 severe stenosis (gradient 100 mm Hg or above). The pulmonary artery electrokymograms were more or less abnormal in all the cases of severe stenosis, in most of those of moderate stenosis, and in one-third of the cases of mild stenosis. Tracings were recorded routinely over the proximal, intermediate, and distal segments of the pulmonary artery

In evaluating the tracings, particular importance was ascribed to the following:

1. Time of onset of upstroke and its relation to the first sound.
2. Occurrence and position of an anacrotic notch and the duration of that part of upstroke preceding the notch, i.e., the initial segment or primary upstroke.
3. Course of upstroke above the notch, i.e., secondary upstroke, and the occurrence of superimposed vibrations.
4. Position of the incisura and shape of the dicrotic wave.

In *mild* stenosis, reduction of the incisura and the dicrotic wave was the only deviation in the shape of the curve (Fig. 157) when the systolic pressure gradient between the right ventricle and the pulmonary artery was less than 30 mm Hg (calculated as the resting value).

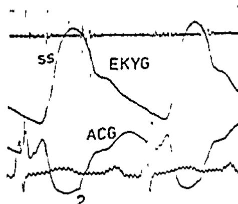


Fig. 157.—Pulmonary artery electrokymogram (EKYG) in mild valvular pulmonary stenosis Woman, aged 20 (T Z 340405). RV pressure 24/1 mm Hg, PA 15/8 Intermediate segment of pulmonary artery PCG over 2nd R I S Curve has normal appearance Low dicrotic wave Systolic click (*ss*) recorded 0 01 sec after start of upstroke ACG apex cardiogram

When the pressure gradient exceeded this figure, a break consistently appeared in systolic upstroke in the form of a halt or notch (Figs. 158 and 159). Its position on the ascending limb of the curve was dependent partly on the grade of stenosis and partly on over which segment of the pulmonary artery the recording was made (Fig. 160) Recorded over the middle seg-

ment. It was generally found at about the middle of the limb, except in the cases in this group with the highest pressure gradi-

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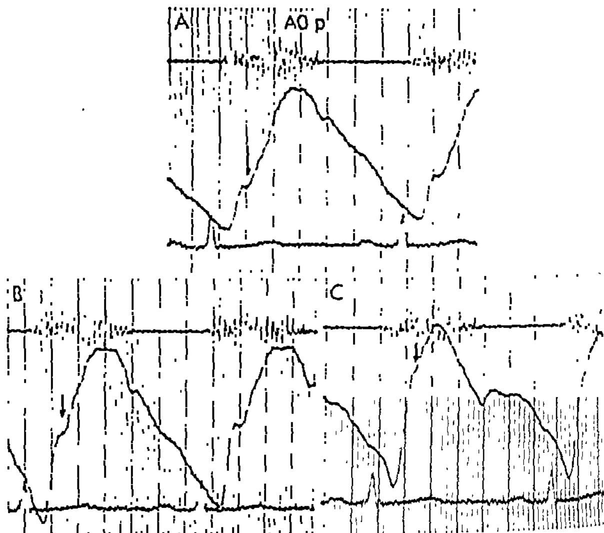


Fig. 160.—Pulmonary artery electrokymograms in mild valvular pulmonary stenosis and atrial septal defect with left to right shunt. Woman, aged 19 (G.S. 360507). RV pressure 50/0 mm Hg, PA 10/3. PCG over 2nd L.I S. Border tracings. *A*, proximal part of pulmonary artery Initial segment starts synchronously with 1st sound and lasts for 0.04 sec, anacrotic notch (arrow) 0 02 sec after beginning of murmur Secondary upstroke has low gradient, summit at level of aortic component of 2nd sound (*AO*); incisura and dicrotic wave high up, 0 02 sec after pulmonary component of 2nd sound (*p*). Relatively low dicrotic wave *B*, intermediate segment of PA. Initial segment has a duration of 0 08 sec, the anacrotic notch (arrow) is high up on systolic limb, 0.06 *sec after* beginning of murmur. Secondary upstroke slightly undulat- [illegible] lower down than in [illegible] nature of rapid sys- [illegible] ura is relatively far down and dicrotic wave distinct.

ment, it was generally found at about the middle of the limb, except in the cases in this group with the highest pressure gradient, in which it was farther down. On the distal tracings, the notch was delayed and appeared higher up on the ascending limb. Multiple small vibrations were often superimposed on the subsequent upstroke, the *gradient of* which decreased with the increase in degree of stenosis. The summit of the curve was round. The incisura and *dicrotic* wave recorded simultaneously with the pulmonary component of the pulmonary second sound were usually diminished Small postdicrotic waves were sometimes present.

In *moderate* stenosis—a systolic pressure gradient between the right ventricle and pulmonary artery of 50–100 mm Hg—the uprise above the anacrotic notch was more continuous and the crest of the curve was peaked (Fig 161, *A*) (This implies a comparison between curves with approximately the same amplitude) In a majority of the studies, the anacrotic notch showed a tendency to be situated in the basal segment of the ascending limb

In *severe* stenosis, in which the pressure gradient amounted to 100 mm Hg or *above*, these features were particularly conspicuous (Figs 161, *B* and 162) The anacrotic notch was invariably present on the curves recorded over the proximal and intermediate levels of the pulmonary artery and occurred at a lower level on the ascending limb than on the curves in the preceding group. *In the most severe cases*, the following upstroke showed a distinct concavity. The incisura and dicrotic wave were absent or were only inappreciable, and then lay far down on the *descending limb*.

The hemodynamic basis of the alterations in the shape of the pulmonary artery pulsations seems to be the following

The stenosed valvular membrane acts, during the later part of the isometric phase of contraction, as a piston which is impelled rapidly toward the pulmonary artery (513: see p 220) and initially dilates it before the ejection phase starts This corresponds on the electrokymogram to the basal part of systolic upstroke (513) and is represented by the initial segment of *the curve*. *Its onset* occurs, on an average, a few hundredths of a second earlier than in normal pulmonary *artery curves*, *starting immediately* before the first heart sound or synchronously with it

On the electrokymogram, this first phase *of dilatation*, which usually lasts for 0 05 to 0.06 sec, is ended by a notch (15). This notch generally appears with a systolic

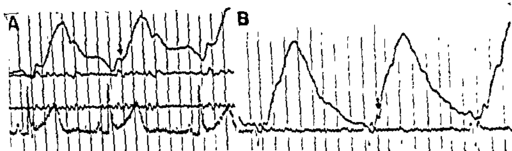


Fig. 161.—A, pulmonary artery electrokymograms in valvular pulmonary stenosis and patent [illegible] 155/0 mm Hg, PA 20/4

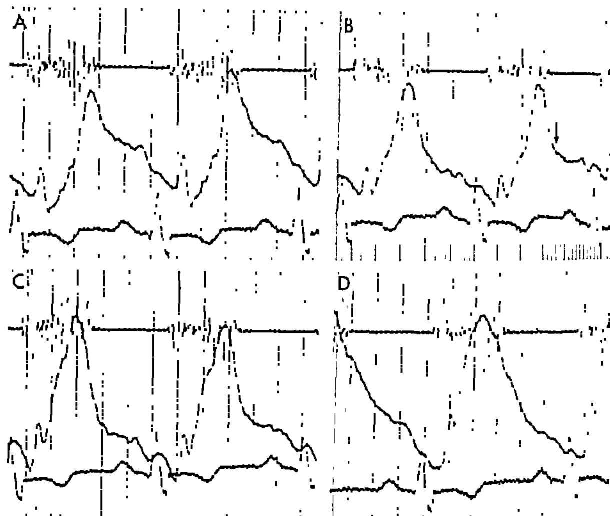


Fig. 162.—Pulmonary artery electrokymograms in severe valvular pulmonary stenosis Man, aged 20 (M.S 350519). RV pressure 170/6 mm Hg, PA 8/4 PCG over 2nd L I S. *A*, presumably very close to pulmonary orifice Start of initial segment synchronous with second major component of 1st sound. Broad "notch," concave ascent of secondary upstroke with superimposed small vibrations, peaked summit late in systole, at end of murmur, and indistinct incisura *B*, distal to *A*. Anacrotic notch is less deep and broad, synchronous with beginning of murmur, summit a few hundredths of second earlier than in *A*. Incisura (arrow) relatively far down *C*, intermediate segment of PA Only upper part of secondary upstroke shows suggested concave course, corresponding to maximum of murmur *D*, distal part of PA Secondary upstroke has a convex course. Summit appears earlier in systole Incisura scarcely demonstrable On all tracings, anacrotic notch lies on lower part of upstroke

pressure gradient across the valve exceeding 30 mm Hg and is presumably a rebound phenomenon, enhanced by the traction effect on the pulmonary artery caused by the downward pull of the pulmonary annulus in ventricular systole (571).

The ascending limb above the anacrotic notch represents the first part of the true ejection phase. The limb starts synchronously with the first vibrations of the systolic murmur (Figs. 160, 162, and 163). During the subsequent rise, the murmur increases in amplitude, and small superimposed waves are often seen on the upstroke They correspond to low-frequency vibrations in the wall of the pulmonary artery, which are plausibly explained as due to turbulence In severe stenosis, the electrokymogram has a tendency to be sinus-shaped (397), which would appear to be an adjustment to the modified type of ventricular contraction (p 185). The concav-

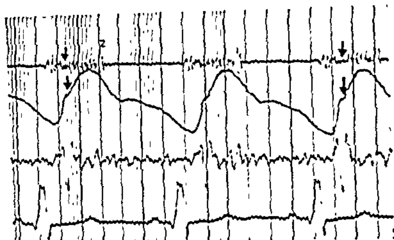


Fig. 163.—Postoperative pulmonary artery electrokymogram in valvular pulmonary stenosis. Boy, aged 10 (C L 440519). PCG over pulmonary area: 2, aortic component of pulmonary 2nd sound, pulmonary component was not recorded Frequency channel, 10 cps. Anacrotic notch (lower arrows) synchronous with the beginning of the systolic murmur (upper arrows). In view of the position of the incisura, closure of the valve should occur 0 08 sec after the aortic component. RV pressure 110/7 mm Hg, PA 18/5.

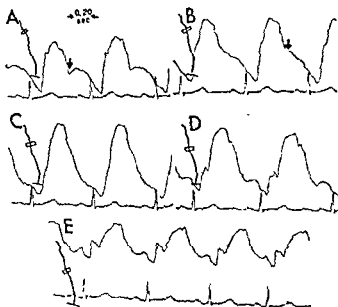


Fig 164 —Pulmonary artery electrokymogram in valvular pulmonary stenosis, showing variations in appearance of the electrokymogram on recording over different segments of the pulmonary artery

ity in the systolic rise observed in very severe stenosis has been interpreted as an expression of retarded expansion of the pulmonary artery due to the low pressure.

Reduction of the incisura and dicrotic wave is a constant feature of the tracings in all degrees of stenosis It presumably represents a decreased rebound resulting from impaired mobility and flexibility of the valve.

One of the most salient features of the electrokymograms in the present series was the variation in shape of the tracings when they were recorded over different levels of the pulmonary artery. As a rule, the greatest pathologic deviations appeared in recordings close to the orifice (Figs 160, 162, and 164) Thus, in tracings recorded basally, the anacrotic notch was broad and deep and situated in the lower part of the ascending limb On recording over the superior part of the pulmonary artery, it ap-

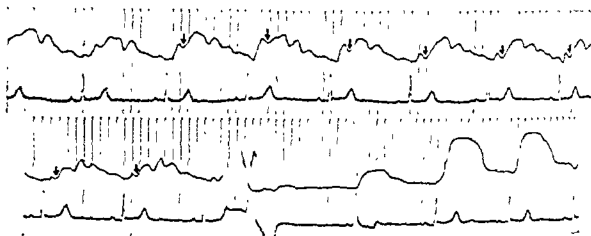


Fig. 165.—Withdrawal curve from the pulmonary artery to right ventricle. Same case as in Figure 166 The *anacrotic notch (arrows) is recorded increasingly far down* on the ascending limb, the closer the catheter tip approaches the valvular plane

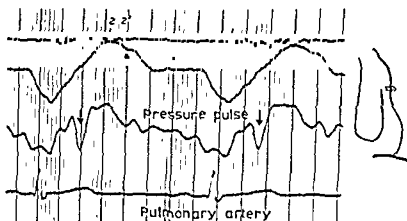


Fig. 166.—Simultaneous recording of electrokymogram and pressure pulse of the pulmonary artery in valvular pulmonary stenosis Girl, aged 10 (E I 440429). Schematic drawing shows position of the catheter tip and photocell PCG over pulmonary area 2 and 2', aortic and pulmonary components of pulmonary 2nd sound Suction effect during maximum ejection (lower arrow), recorded synchronously with the greatest amplitude of the murmur, is not reflected on the electrokymogram, consequently, inverse pulsations in the pulmonary artery cannot be demonstrated The incisura on the electrokymogram (upper arrow) and pressure curve are recorded simultaneously with the pulmonary component of the pulmonary 2nd sound RV pressure 45/5 mm Hg, PA 8/2.

peared higher up on the curve and was less prominent. It is interesting that this variation in position of the anacrotic notch had, in some cases, its equivalent in appearance of the pressure pulse curve at varying distances from the pulmonary valve (Fig. 165). As far as the incisura and dicrotic wave are concerned, the reverse applied; they were most distinct in tracings from the superior segment of the artery. As under normal conditions (p. 111), they were inconspicuous on the tracings from the basal part of the artery. On the other hand, an exact correspondence to the pulmonary component of the pulmonary second sound was found in these curves. The gradient of the secondary part of the systolic upstroke, above the notch, was low in the tracings made basally, whereas it increased in those recorded more superiorly.

Exaggeration of the abnormal features on the tracings recorded close to the valve was ascribed to the fact that the hemodynamic changes are most marked in this part of the artery and are reflected distinctly on the electrokymograms

Density electrokymograms were essentially in agreement with the curves recorded over the border of the pulmonary artery Similar tracings could be recorded over the vessels of the hilum.

Attempts were also made to record elec-
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... of the jet of blood from the orifice No special change in the shape of the electrokymogram produced by the jet could be demonstrated Superimposition of the aorta hampered the investigation

An anacrotic notch—which is unquestionably the most characteristic feature of the pulmonary artery electrokymogram in pulmonary stenosis—can, however, be observed in other conditions, e g mitral stenosis and sometimes pulmonary hypertension But in these conditions, the incisura and dicrotic notch are not usually delayed, and the consistent variation ...
...

artery is not as typical. There are also other differences, which are dependent on the time of onset of the systolic upstroke and on the size of the incisura and dicrotic wave. In both pulmonary hypertension and severe pulmonary stenosis, the electrokymogram is characterized by a tendency to a sinus shape. In the former, the incisura and dicrotic wave are situated high up on the descending limb (see p. 365), but in severe pulmonary stenosis are lower down.

An important source of error arises when the recording is made too far down, with

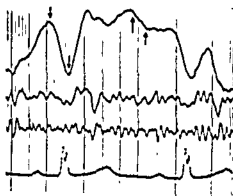


Fig. 167.—Right atrial electrokymogram in moderately severe valvular pulmonary stenosis Boy, aged 11 (L.W. 421111). Atrial contraction (between left hand arrows) lasts 0 10 sec and is deep in relation to the emptying phase during early diastole (between right-hand arrows), indicating impediment to emptying of atrium RA pressure, *a* wave 10 mm Hg, mean pressure, 4 mm

the slit placed partly over the auricular appendage. A superimposed wave from the left atrium then appears in the upstroke of the pulmonary artery curve, simulating the presence of an anacrotic wave. It may also be recalled that an anacrotic notch, which marks the end of the isometric phase of contraction in the right ventricle, can normally be recorded over the most basal segment of the pulmonary artery.

The impediment to emptying of the *right atrium* in moderately severe and severe pulmonary stenosis is seen on fluoroscopy ...

enlarged. On the electrokymogram (Figs. 167 and 168) it is recorded as a prolonged and deepened presystolic deflection (16). In our cases, the abnormal atrial contraction generally lasted for 0.10 sec or more. In a few cases with unquestionably impeded emptying, it was shorter, but it was invariably deep in relation to the emptying phase in early diastole. When the pulsations were recorded with the patient in the recumbent position, the duration of contraction increased (see Fig. 169), as is almost invariably the case under normal conditions.

Supravalvular Stenosis

In this case as well, alterations were present in the electrokymograms from the pulmonary artery and resembled those in valvular stenosis. A notch occurred in the systolic uprise, and the incisura and dicrotic wave were reduced.

Infundibular Stenosis

Alterations were present in the electrokymograms of the pulmonary artery in 15 cases of infundibular stenosis, including

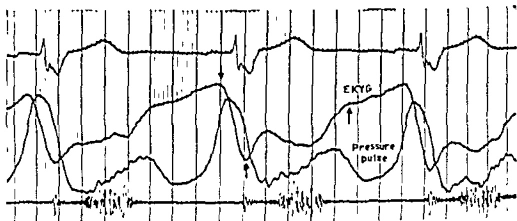


Fig. 168.—Right atrial electrokymogram and pressure pulse in severe valvular pulmonary stenosis. Girl, aged 10 (B.L. 440127). PCG over (between left-hand arrows) 0.11 sec, it is deep, recorded only as an inflection (right-hand arrow of an impediment to atrial emptying Atrial contraction occurs simultaneously with peak of the a wave, which measures 10 mm Hg

Curves of the same type as in the form of impaired emptying of the atrium just described can be recorded in other conditions, e g., primary pulmonary hypertension, constrictive pericarditis, and heart failure (475, 571). Consequently, the electrokymogram gives no information regarding the nature of the impediment.

A few cases with an associated right to left interatrial shunt were also studied. The electrokymographic records of both the right and the left atrium had a normal appearance.

In tracings made over the infundibulum of the right ventricle, the onset of the ejection phase was sometimes well defined (Fig. 170).

the simple type and that with coincident ventricular septal defect with or without a shunt. The appearance of the tracings differed from that typical of valvular stenosis Normal curves were recorded in five cases of mild infundibular stenosis with a pressure gradient across the pulmonary valve not exceeding 30 mm Hg.

Characteristic features of the pulmonary artery curves were a late incisura and dicrotic wave and a more or less horizontal course in systole. The curves thus lacked any real summit and no sinus shape was present (Figs. 171-173). The incisura coincided with the pulmonary component of the second sound on the phonocardiogram. The upstroke was sometimes early and had

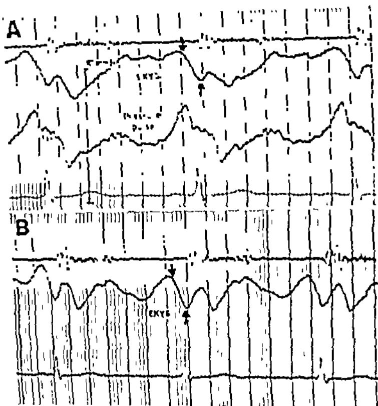


Fig 169 —Right atrial electrokymograms and pressure pulse in mild to moderate [illegible]

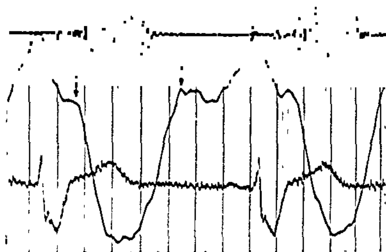


Fig. 170.—Electrokymogram of infundibulum in severe valvular pulmonary stenosis Girl, aged 10 (B L. 440127). PCG over pulmonary area. Recording made in true lateral projection. At the start of the emptying phase, synchronously with the first vibrations of the murmur, a halt (left-hand arrow) occurs in the systolic deflection. The peak (right-hand arrow) in the ascending limb coincides approximately with closure of the valve.

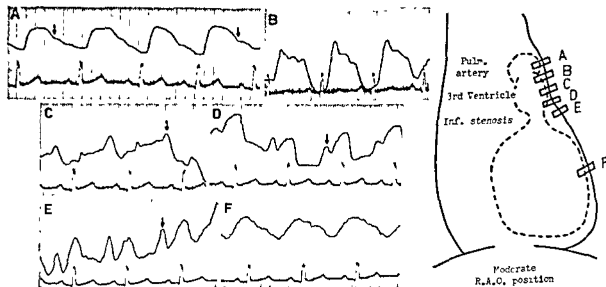


Fig. 171 (*left*) —Pulmonary artery and right ventricular electrokymograms in infundibular stenosis and associated ventricular septal defect, recorded over segments shown in schematic drawing (*Fig 172*). *Girl, aged 6 (M H 470717) A, pulmonary artery, plateau* shape during systole, late incisura (arrow) and reduced dicrotic wave *B*, superior part of infundibulum: the tracing shows the expansion during systole *C–E*, farther down over the third ventricle: an inward movement of the wall is recorded during systole, a peak (arrows) marks transit into the relaxation phase *F*, right ventricle below the stenosis ordinary tracing RV pressure 115/9 mm Hg, third ventricle 12/7, PA 12/2

Fig. 172 (*right*) —Schematic representation of position of the photocell in case illustrated in Figure 171.

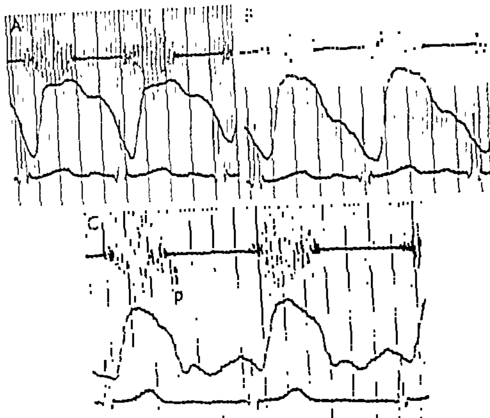


Fig 173.—Pulmonary artery electrokymograms in infundibular stenosis with intact ventricular septum. Girl, aged 8 (M H. 470414). PCG over 2nd L I S : *p*, pulmonary component of 2nd sound *A*, proximal segment Plateau-shaped course, small superimposed vibrations in systole, and suggested concavity at maximum of murmur. *B*, distal segment of PA. Distinct summit in systole. Onset of descending limb at end of T wave, reduced dicrotic wave. Upstroke at first major component of 1st sound. C, presumably at border of infundibulum Upstroke before 1st heart sound Fall in later part of systole

a high gradient. The abnormalities in the curves appeared chiefly on tracings recorded over the proximal segment of the artery, whereas those recorded distally sometimes had a normal configuration.

An anacrotic notch was present in several cases with associated valvular stenosis (Fig. 174). Contrary to the conditions in valvular *stenosis alone, the rise above the* notch was steepest in the electrokymograms recorded over the basal segment of the artery.

An anacrotic notch was also observed in one case of purely infundibular stenosis. The stenosis was severe. The notch appeared on the tracings from both the pulmonary artery and the infundibulum. The lower part of the rise in these curves may be due to an impact from the infundibular stenosis, which would explain the presence of the notch in this case.

In several cases, the findings at angiocardiography were utilized to facilitate the placing of the photocell for a study of the pulsations of the third ventricle The tracings from the superior part of the infundibulum showed systolic expansion due to influence of the *overlapping pulmonary ar*tery; elsewhere—including the stenosed area—the contraction appeared on the record (Figs. 171 and 172). Below the stenosis, the tracings had a normal appearance.

The electrokymogram of the right atrium was characterized in two cases of severe stenosis by increased presystolic activity as a manifestation of an impediment to emptying (Fig. 175).

CARDIAC CATHETERIZATION

As we have already mentioned, it is only since cardiac catheterization was introduced as a clinical method that pulmonary stenosis with normal aortic root has become a generally recognized clinical entity. Our knowledge of the characteristic clinical picture has been enhanced by this means, so that it is now usually possible to diagnose the condition even without catheterization. It is, however, necessary to resort to cardiac catheterization for an exact determination of the degree of stenosis This examination was performed in all patients of our series

The width of the orifice can be calculated if the pressure gradient between the right *ventricle and the pulmonary artery,* the

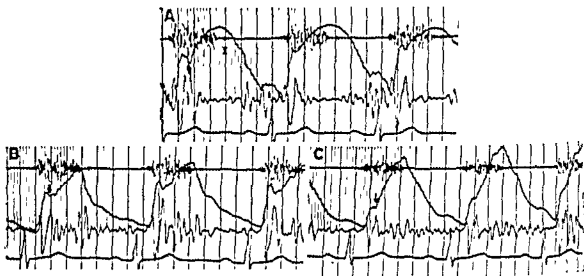


Fig. 174.—Pulmonary artery electrokymograms in combined valvular and infundibular stenosis and ventricular septal defect *Boy, aged 10 (C F 440603) PCG over pulmonary area* Frequency channel, 15 cps *A*, superior, *B*, middle, and *C*, inferior segment of pulmonary artery Arrows point to anacrotic notch, steepest gradient of the upstroke above the notch is recorded in the tracings made over the inferior segment Incisura and dicrotic wave are reduced

stroke volume, the ejection time, and the viscosity of the blood are known. If the patient is in a relatively basal condition at examination, the systolic pressure in the right ventricle can be used as a gauge of the degree of stenosis In the severest stenoses, the pulmonary flow may be diminished (high arteriovenous oxygen difference), and the stenosis is therefore more

these cases to enter the pulmonary vein and to measure the pulmonary venous wedge (PCA) pressure. In some of the severest cases of stenosis, we made no attempt to pass the catheter through the pulmonary orifice, in view of the risk of arresting the flow completely.

When the pressure in the main trunk of the pulmonary artery is recorded in a pa-



Fig. 175.—Right atrial electrokymogram. Same case as in Figure 174. PCG over pulmonary area Frequency channel, 15 cps. Atrial contraction (between arrows) prolonged (duration 0 10 sec); it is deep in relation to emptying phase in early diastole

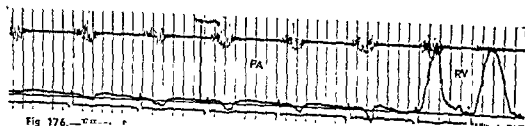


Fig 176.— ... 481127) On p ... tion of flow, a ... catheter tip in ... pressure in the ... phy shows that the

severe than is indicated by the pressure in the right ventricle.

The systolic pressure in the right ventricle in our series can be inferred from Figure 129 (p 143). The lowest pressure recorded was 31 *mm* Hg and the highest 270 mm Hg The smallest pressure gradient across the pulmonary valve was 14 mm Hg. The pressure in the pulmonary artery was normal ... in four of

tient with severe pulmonary stenosis, the value obtained is lower than the actual value. This is dependent on the following two factors

1. The catheter causes additional narrowing of the orifice. Consequently, in all such cases the patient must be observed closely when the catheter passes the orifice. In one of our cases, the orifice became completely occluded and no pulsations from the pulmonary artery could be recorded. No peripheral pulse could be palpated The catheter was immediately withdrawn into

the ventricle before the patient had suffered any untoward effect.

2. The pressure recorded in the pulmonary artery is influenced by the high velocity of flow and will therefore be *too low* when the opening of the catheter lies in the direction of the flow. The rate of flow is greatest at the site of the stenosis and immediately distal to it. This can be observed on recording of the pressure when the catheter is withdrawn from the pulmonary artery into the ventricle. Figure 176 shows a

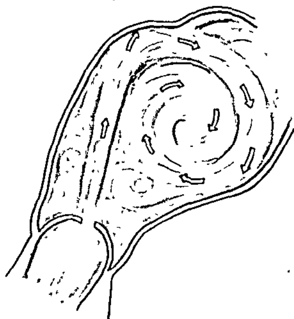


Fig. 177.—Schematic drawing of valvular stenosis with poststenotic dilatation. Eddies arise in the main trunk of the pulmonary artery, pressure therefore varies in the different parts of the vessel

withdrawal curve of this nature in a patient with severe valvular pulmonary stenosis without ventricular septal defect No effect of the pressure of velocity can be noted in the peripheral part of the pulmonary artery, but the nearer the catheter tip approaches the pulmonary orifice, the greater the fall in pressure during maximum ejection

The phonocardiogram showed that this fall in pressure occurred when the systolic murmur was maximal. The area of the orifice was determined approximately on the basis of the angiocardiogram. The pressure of velocity was calculated to be about 40 mm Hg. Bayer *et al.* (45) published a curve with a similar appearance, and they also stressed the importance of the pressure of velocity. They stated that this type of curve is characteristic of severe infundibular stenosis. They did not mention how the anatomic diagnosis of the stenosis was confirmed. In our opinion, this pressure tracing is more characteristic of pure valvular stenosis. Obviously, the pressure of velocity affects the pressure curve in infundibular stenosis as well, but not to as great an extent, since severe infundibular stenosis is almost invariably associated with a ventricular septal defect. Consequently, the rate of flow is never as high as in severe valvular stenosis.

In valvular stenosis, a poststenotic dilatation takes place in the main trunk of the pulmonary artery. It is partly dependent on the turbulence arising on the rapid flow through the stenosis (Fig. 177). The pressure therefore varies considerably in this vascular section (adjacent to the valvular plane). An entirely different pressure would be obtained if it were recorded beside the vessel wall instead of centrally at the orifice. As a rule, the relatively inflexible catheter lies in the center of the blood flow, and the pressure of velocity comes into effect.

On the basis of the appearance of the withdrawal curve, it is possible to distinguish between valvular, infundibular, and combined stenosis This has been stressed in the majority of publications on the hemodynamics of pulmonary stenosis For this purpose, it is essential to move the catheter extremely slowly, under fluoroscopic control and simultaneous pressure recording

Withdrawal curves of this kind in various types of pulmonary stenosis are shown in Figure 178 The anatomic diagnosis was confirmed in every instance by angiocardiography. In valvular stenosis, there is a sudden transition from pulmonary to ventricular pressure. The pressure is sometimes lowered slightly distal to the valve, owing to the pressure of velocity In circumscribed, infundibular stenosis with a third ventricle, an intermediate zone is obtained,

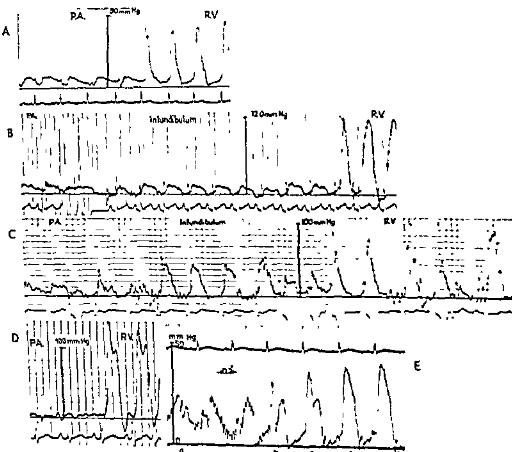


Fig 178 —Withdrawal ...
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its systolic pressure is the same as that in the pulmonary artery, whereas the diastolic pressure is that of the ventricle and the shape of the curve is of the ventricular type. *In combined valvular and infundibular* stenosis, systolic pressure in the intermediate zone is higher than that in the pulmonary artery but lower than that in the *right* ventricle. *When the* infundibular stenosis forms a long, constricted channel—seen only in the tetralogy of Fallot—no intermediate zone can be observed.

The sources of error of this method have not always been taken into account. There has not infrequently been failure to confirm the anatomic diagnosis with other methods of investigation. Kirklin *et al.* (389) nevertheless point out that a sudden transition from arterial to ventricular pressure can be observed not only in valvular stenosis but also in purely infundibular stenosis if it is situated close to the valve, as well as in valvular stenosis combined with extremely slight infundibular stricture. In our experience, too, it may be impossible to distinguish valvular from infundibular stenosis of channel type or with only a small third ventricle This applied in two of our cases, in which the withdrawal curve had the same shape as in valvular stenosis, but the angiocardiogram showed the presence of infundibular stenosis only It often is difficult to interpret the withdrawal curve, owing to the numerous ventricular extrasystoles occurring when the catheter passes through the infundibulum. If, in valvular *stenosis, the catheter tip lies in the orifice,* it may pass into the pulmonary artery during systole and slip back into the right ventricle during diastole. The pressure curve *may then resemble that obtained from the* third ventricle in infundibular stenosis (Fig. 178, *E*)

In this connection, it is important to recall that in severe valvular stenosis there is great hypertrophy of the muscle wall in the infundibulum In the opinion of some authors (71, 95, 336, 389), this hypertrophy may be sufficient to produce an infundibular stenosis. At autopsy, the infundibulum certainly presents as a long canal, but in *our cases studied* by angiocardiography, no true stenosis was found. It is only at the end of systole, after the phase of maximum ejection and in the beginning of diastole, *that a strong contraction* is observed which causes considerable narrowing of the infundibulum (see p. 220). Thus, this muscular hypertrophy constitutes no obstacle to emptying of the right ventricle, and it is therefore incorrect—as Bing *et al.* (57) have also stressed—to speak of infundibular stenosis. Consequently, we consider infundibular resection to be contraindicated in cases of this nature. In five such cases of valvular stenosis, in which angiocardiography was performed pre- and postoperatively, we found appreciable widening of the infundibulum five to 12 months after valvulotomy (see Fig. 187, p. 195). A true malformation of the infundibulum, with stenosis of the ostium infundibuli combined with valvular pulmonary stenosis, which is so common in tetralogy of Fallot, is uncommon when the ventricular septum is intact

After operation for valvular *stenosis,* several authors have found a persistent pressure gradient between the right ventricle and pulmonary artery, which has been considered to be due to hypertrophy *of the* infundibulum (71, 95, 336). In our experience, a persistent pressure gradient is to be ascribed to incomplete valvulotomy. A pressure gradient *between the right* ventricle and pulmonary artery in excess of 20 mm Hg was recorded in eight of 23 cases in which catheterization was performed postoperatively. *In all these cases* the withdrawal curve had the appearance seen in valvular stenosis, and in a few cases in which angiocardiographic examination was *carried out, it showed* persisting valvular stenosis These cases will be reported *in detail* by Hanson *et al* (318).

When the site of stenosis is judged only on the basis of the appearance of the withdrawal curve, it is necessary to *avoid the* sources of error discussed on this page (Fig 178, E). Certain of the pressure curves which, in this connection, have been published as evidence of the presence of infundibular stenosis have exhibited the fea-

tures seen in valvular stenosis if, during recording, the catheter tip moved between the pulmonary artery (in systole) and the right ventricle (in diastole).

In circumscribed infundibular stenosis, the infundibulum forms a third ventricle of varying size. The dynamic course in this chamber differs essentially from that in the remainder of the right ventricle, and this does not apply only to the level of the systolic pressure. As a rule, infundibular stenosis is combined with ventricular septal defect. If stenosis is only moderate, a left to right shunt develops (p. 138).

Because cases with an increased flow of maximum ejection. The systolic contraction persists until 0.46 sec after the R wave, i e., relaxation of the third ventricle takes place 0.08 sec later than in the inflow tract. The pressure in the pulmonary artery does not start to rise until 0 08 sec after the R wave, and, here as well, the pressure of velocity results in a slight fall in pressure. A notch on the descending limb 0.46 sec after the R wave indicates closure of the pulmonary valve. A study of the phonocardiogram discloses that the aortic component of the second heart sound coincides with the end of systole in the inflow tract of the right ventricle and that the pulmo-

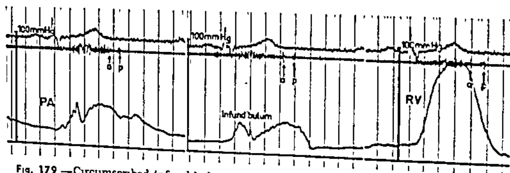


Fig. 179 —Circumscribed infundibular stenosis with ventricular septal defect Pressure recordings from main trunk of the pulmonary artery (PA), infundibulum and inflow ... ventricle (RV) simultaneous record ... right ventric ... the infundib ... sound (p) ar ... component c ... through the ... marked and ... onary artery.

through the infundibulum exhibit more marked changes, we have chosen one such case to illustrate the hemodynamics in circumscribed infundibular pulmonary stenosis Figure 179 shows the pressure recordings in the pulmonary artery and the third and right ventricles The electrocardiogram and phonocardiogram were recorded simultaneously The isometric contraction in the inflow tract of the right ventricle is seen to start 0 05 sec and systole to end 0 38 sec after the peak of the R wave The pressure in the third ventricle rises simultaneously with that in the inflow tract, but a fall in pressure caused by the pressure of velocity occurs during the phase

nary component coincides with the end of systole in the third ventricle and the notch in the pulmonary artery ... murmur ... of the se... in the ma... part of the right ventricle), i e., when the blood flow through the infundibular stenosis has ceased Since no valvular stenosis is present, emptying of the third ventricle is not associated with a murmur. Nor is a murmur recorded between the aortic and pulmonary components of the second heart sound (see p. 149).

In severe pulmonary stenosis without ventricular septal defect, there is a change in the appearance of the ventricular curve

Wiggers (701) found experimentally a change in the shape of the curve in connection with progressive circular constriction of the pulmonary artery. The shape of the ventricular curve was typical of an isometric mode of contraction. The duration of systole was unaffected. A rise in pressure occurred at the end of diastole. Such experimental observations cannot be applied without reservation to the hemodynamics in pulmonary stenosis, in which there is considerable muscular hypertrophy and the pressure in the right ventricle is raised to a far greater degree than in these experiments.

An example of a ventricular pressure curve in pulmonary stenosis is shown in Figure 178 Its contour is seen to be peaked The summit of pressure is reached late in systole. This is also evident from Figure 180, in which a comparison is made of the recordings in 18 of the patients with the most severe stenosis and in 14 healthy individuals. In acute experiments Wiggers found, on the contrary, that the greater the degree of stenosis, the earlier the summit of pressure was reached.

The pressure in the right atrium also undergoes a change in *severe* pulmonary stenosis with intact ventricular septum The hypertrophied right ventricle presents greater resistance than normally to diastolic filling. The filling pressures rises This elevated pressure in the right atrium is characterized in particular by a large presystolic rise in pressure (a tall, peaked *a* wave, which also appears on the venous pulse curve) (2). Figure 182, *A*, shows the pressure in the right atrium in a patient with systolic pressure in the right ventricle of 230 mm Hg The correlation between the height of the *a* wave and the systolic pressure in the right ventricle can be inferred from Figure 181.

In order to make a complete diagnosis in these cases, it does not suffice to demonstrate the presence of pulmonary stenosis and to determine its degree. It is essential, in addition, to ascertain whether a septal defect is present and then to analyze the

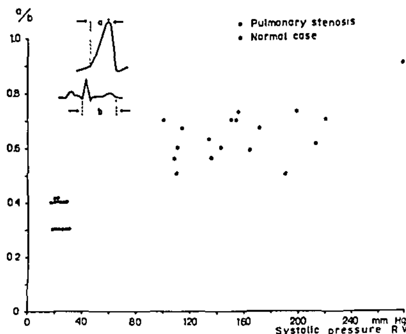


Fig. 180.—Time relation of peak of right ventricular pressure to R–T interval (*a* to *b*) in normal cases and most severe cases of pulmonary stenosis. Correlation with systolic pressure in right ventricle Peak of right ventricular pressure occurs later in systole in severe pulmonary stenosis than in normal cases Interval from beginning of ventricular systole to its peak, *a*, R–T interval, *b*

shunt A left to right shunt can be demonstrated on catheterization of the right side of the heart. In our series we were able in this way to detect seven cases of an atrial septal defect and 14 of a ventricular septal defect; in view of the left to right shunt, we classified them as septal defects

The presence of a right to left shunt is established by means of arterial puncture. In severe pulmonary stenosis combined with ventricular septal defect, the right to left shunt may be large. Such cases were classified as tetralogy of Fallot even if the the shunt is correlated to the degree of stenosis and width of the foramen ovale.

The combination of pulmonary stenosis and ventricular septal defect without a shunt produces the same clinical features as in intact ventricular septum. Catheterization shows the systolic pressure in the right ventricle to lie on the same level as that in the left ventricle or the systemic circulation. It must, however, be borne in mind that the systolic pressure in a peripheral systemic artery is normally somewhat higher than that in the left ventricle. But

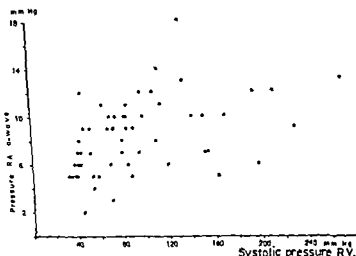


Fig. 181.—Correlation between systolic pressure in right ventricle and presystolic rise in pressure in right atrium (level of *a* wave) in pulmonary stenosis with intact ventricular septum Raised pressure in right atrium is present in most cases of severe stenosis but may also be found in mild cases

aorta was not over-riding. In some cases the stenosis is of such degree that a shunt cannot be demonstrated at rest but only on exertion In severe stenosis with intact ventricular septum, there may be a right to left shunt through an atrial septal defect or a [illegible]

more severe the pulmonary stenosis, the more marked is the rise in pressure in the right atrium, especially during atrial systole By means of a comparison between the clinical and the anatomic picture, Selzer and Carnes (591) showed that the size of

even in pulmonary stenosis with intact ventricular septum, the systolic pressure in the right ventricle is often found to be on the level of the systemic pressure (e.g. Case 25/57 in Table 4). Consequently, in older children and adults, an exercise tolerance test may be of value for a differential diagnosis. Thus it should be possible to rule out a ventricular septal defect in Case 46/57 (Table 4), since the pressure in the right ventricle rose from 105 to 187 mm Hg without any fall in arterial oxygen saturation, despite a simultaneous rise in pressure in the brachial artery from 121 to 154 mm Hg. This is, however, necessary with a heavy

TABLE 4.—Hemodynamics at Rest and During Exercise in Five Adult Patients with Pulmonary Stenosis of Different Types*

Case No	Sex	Age Year	Body Surface Area, M²	Physical Working Capacity % of Predicted	Work Load kgm/min	O₂ Uptake, ml/min	Pulse Rate, beats/min	Pulmonary Circulation: AV O₂ Diff, ml/L	Pulmonary Circulation: Cardiac Output L/min	Pulmonary Circulation: Stroke Vol, ml	Systemic Circulation: AV O₂ Diff, ml/L	Systemic Circulation: Cardiac Output, L/min	Systemic Circulation: Stroke Vol, ml	O₂ Cap vol %	Art O₂ Sat, %	Shunt, L/min: R → L†	Shunt, L/min: L → R	Pressure, mm Hg: RA Mean	Pressure, mm Hg: RV Syst	Pressure, mm Hg: PA Syst	Pressure, mm Hg: PA Diast	Pressure, mm Hg: PA Mean	Pressure, mm Hg: Br. A Syst	Pressure, mm Hg: Br. A Diast	Pressure, mm Hg: Br. A Mean	RV Systolic Duration sec×100	Anatomy
6/55	M	17	1.62	107	Rest	258	84	33	7.8	93	—	—	—	18.1	96	—	—	4	35	17	8	12	115	61	—	38	Valvular stenosis
					300	828	112	86	9.6	86	—	—	—	18.5	96	—	—	—	51	—	—	19	145	79	—	32	
					600	1467	172	123	11.9	69	—	—	—	19.0	96	—	—	—	76	32	8	18	162	100	—	24	
46/57	M	29	1.96	83	Rest	327	84	36	9.1	109	—	—	—	17.6	98	—	—	8	105	19	5	11	121	69	87	40	Valvular stenosis
					300	923	126	76	12.2	97	—	—	—	17.6	98	—	—	10	157	27	4	15	148	74	96	32	
					600	1410	159	92	15.3	96	—	—	—	18.1	96	—	—	10	187	27	5	15	154	72	96	26	
56/56	F	25	1.53	10	Rest	203	90	115	1.8	20	58	3.5	39	19.7	69	1.7	—	7	220	—	—	8	109	67	83	40	Valvular stenosis + atrial septal defect
					50‡	—	136	—	—	—	—	—	—	—	51	—	—	—	220	—	—	—	142	86	—	37	
25/57	M	24	1.85	27	Rest	332	84	86	3.9	46	50	6.7	80	26.1	83	2.8	—	—	146	12	4	7	145	87	106	—	Valvular stenosis + atrial septal defect
					200	764	123	169	4.5	37	72	10.6	86	26.2	58	6.1	—	—	—	15	3	7	157	81	106	—	
					400	1096	165	218	5.0	30	64	17.2	104	27.0	40	12.2	—	—	—	22	0	13	187	89	120	—	
3/56	M	25	1.94	82	Rest	306	62	28	10.9	176	33	9.3	150	17.8	95	—	1.6	5	73	28	7	11	110	67	80	42	Infundibular stenosis + ventricular septal defect
					300	867	98	75	11.6	118	68	12.8	131	18.0	89	1.9	0.7	5	102	31	5	16	136	80	98	38	
					600	1558	152	132	11.8	78	61	25.5	168	18.6	58	13.7	—	—	130	37	—	15	142	73	98	27	

*For abbreviations see Table 1, p. 119. †Pulmonary venous blood assumed to be 97 per cent saturated. ‡Only 2½ min of work. Cardiac output not determined.

load of work Since the systolic pressure in the systemic circulation normally rises on exertion (344), the pressure in the right ventricle may, even with an intact septum, coincide with the systemic pressure both at rest and on mild exertion

A ventricular septal defect can be demonstrated by passage of the catheter into the aorta, which often is possible in overriding aorta but seldom in ventricular septal defect with normal aortic root. It was, however, possible in six of our cases.

The most certain way of ruling out a ventricular septal defect in these cases is by angiocardiography with injection of contrast medium into the right ventricle or possibly into the left. Even if no right to left shunt exists in the presence of a ventricular septal defect, the contrast medium injected into the right ventricle will be forced into the left ventricle *during diastole* if the injection is made rapidly. Many authors are of the opinion that a septal defect cannot be demonstrated with certainty by angiocardiography (502, 676). We performed this examination in 68 cases of pulmonary stenosis and were able in all of them not only to ascertain the anatomy of the stenosis, but also to demonstrate the presence of any existing defects of the ventricular septum

The differentiation between tetralogy of Fallot and pulmonary stenosis with intact ventric...

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pre ...ulties The physical findings, electrocardiogram, phonocardiogram, and roentgenologic examination often provide sufficient guidance

At catheterization, a right to left interatrial shunt can be demonstrated by passage of the catheter through the atrial communication, which is nearly always successful if the catheter is inserted through a leg vein An interatrial shunt does not, however, *rule* out the existence of a ventricular septal defect as well As a rule, the communication between the atria consists of a patent foramen ovale covered by a valve, but a true atrial septal defect may be present (132). After valvulotomy, the right to left shunt gradually disappears, and no left to right shunt arises if the foramen ovale has been covered by a valve. An atrial septal defect can often be easily distinguished from a patent foramen ovale with the balloon catheter technique (see p 445). This simple method was used in six of our cases, a patent foramen ovale could be demonstrated in four of them and a true atrial septal defect in two

The position of the shunt can be determined by studying the dilution curve or circulation time after injection of dye, ether, or fluorescein into the right atrium and right ventricle (46, 128, 409, 723). However, if some of the test substance injected into the right ventricle regurgitates into the atrium, the examination may be misleading.

In cases with an intact septum and an interatrial shunt, the systolic pressure in the right ventricle as a rule is considerably higher than that in the systemic circulation. That this does not invariably apply is shown by Case 25 57 (Table 4). Unfortunately, the ventricular pressure during exercise was not recorded, but probably it would not have risen more than that in the brachial artery.

In these cases as well, we have found angiocardiography to be the method of choice for establishing the anatomy of the malformation in detail. Sometimes, in fact, this examination is entirely decisive *for a differential diagnosis* between tetralogy of Fallot and pulmonary stenosis with intact ventricular septum and interatrial shunt.

The anatomic diagnoses in our series of cases are recorded in Table 3 (p 141) Of the 13 patients with a ventricular septal defect, only five had decreased arterial oxygen saturation at rest (89, 69, 87, 67, and 86 per cent, respectively). In three of them it was also possible *to* pass the catheter through the patent foramen ovale into the left atrium, in which the oxygen saturation was normal

A right to left interatrial shunt was present in nine cases The arterial oxygen saturation ranged from 64 to 92 per cent, It

TABLE 4.—Hemodynamics at Rest and During Exercise in Five Adult Patients with Pulmonary Stenosis of Different Types*

Case No	Sex	Age, Year	Body Surface Area, M²	Physical Working Capacity, % of Predicted	Work Load kgm/min	O₂ Uptake, ml/min	Pulse Rate, beats/min	Pulmonary Circulation: AV O₂ Diff, ml/L	Pulmonary Circulation: Cardiac Output L/min	Pulmonary Circulation: Stroke Vol, ml	Systemic Circulation: AV O₂ Diff, ml/L	Systemic Circulation: Cardiac Output, L/min	Systemic Circulation: Stroke Vol, ml	O₂ Cap vol %	Art O₂ Sat, %	Shunt, L/min: R → L†	Shunt, L/min: L → R	Pressure, mm Hg: RA Mean	Pressure, mm Hg: RV Syst	Pressure, mm Hg: PA Syst	Pressure, mm Hg: PA Diast	Pressure, mm Hg: PA Mean	Pressure, mm Hg: Br. A Syst.	Pressure, mm Hg: Br. A Diast.	Pressure, mm Hg: Br. A Mean	RV Systolic Duration sec × 100	Anatomy
6/55	M	17	1.62	107	Rest	258	84	33	7.8	93	—	—	—	18.1	96	—	—	4	35	17	8	12	115	61	—	38	Valvular stenosis
					300	828	112	86	9.6	86	—	—	—	18.5	96	—	—	—	51	—	—	19	115	79	—	32	
					600	1467	172	123	11.9	69	—	—	—	19.0	96	—	—	—	76	32	8	18	162	100	—	24	
46/57	M	29	1.96	83	Rest	327	84	36	9.1	109	—	—	—	17.6	98	—	—	8	105	19	5	11	121	69	87	40	Valvular stenosis
					300	923	126	76	12.2	97	—	—	—	17.6	98	—	—	10	157	27	4	15	148	71	96	32	
					600	1410	159	92	15.3	96	—	—	—	18.1	96	—	—	10	187	27	5	15	151	72	96	26	
56/56	F	25	1.53	10	Rest	203	90	11.5	1.8	20	58	3.5	39	19.7	69	1.7	—	7	220	—	—	8	109	67	83	40	Valvular stenosis + atrial septal defect
					50‡	—	136	—	—	—	—	—	—	—	54	—	—	—	220	—	—	—	142	86	—	37	
25/57	M	24	1.85	27	Rest	332	84	86	3.9	46	50	6.7	80	26.1	83	2.8	—	—	146	12	4	7	145	87	106	—	Valvular stenosis + atrial septal defect
					200	764	123	169	4.5	37	72	10.6	86	26.2	58	6.1	—	—	—	15	3	7	157	84	106	—	
					400	1096	165	218	5.0	30	64	17.2	104	27.0	40	12.2	—	—	—	22	0	13	187	89	120	—	
3/56	M	25	1.94	82	Rest	306	62	28	10.9	176	33	9.3	150	17.8	95	—	1.6	5	73	28	7	11	110	67	80	42	Infundibular stenosis + ventricular septal defect
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					600	1558	152	132	11.8	78	61	25.5	168	18.6	58	13.7	—	—	130	37	—	15	142	73	98	27	

*For abbreviations see Table 1, p 119 †Pulmonary venous blood assumed to be 97 per cent saturated ‡Only 2½ min of work Cardiac output not determined

cal picture in agreement with that in tetralogy of Fallot. During exercise (Case 25/57) the stroke volume decreased in the right ventricle at the same time that it increased in the left ventricle; i.e., the right to left shunt increased, so that the arterial oxygen saturation fell from 83 to 40 per cent. The mechanical systole in the right ventricle could be determined in only one of the cases (56/56). The working capacity was so low that the patient was able to work only for two and one-half minutes with a very low load. The pulse rate then rose to 136 beats per minute, and the arterial oxygen saturation fell from 69 to 54 per cent. The stroke volume at work could not be determined. It was, however, evident that the right ventricle was unable to raise the pressure and shorten mechanical systole to the normal extent (see Fig. 183). As a result, the shortening of diastole at a high pulse frequency was so considerable that diastolic filling of the ventricle decreased.

In pulmonary stenosis combined with ventricular septal defect, the direction and size of the shunt vary with the load of work (Case 3/56). This shows the difficulty of classifying such cases. In the case in question the left to right shunt at rest was, however, so small that the clinical picture was identical with that in pulmonary stenosis with intact ventricular septum.

ANGIOCARDIOGRAPHY

Valvular Stenosis

From the surgical point of view, it is desirable for the angiocardiographic examination to be aimed mainly at elucidating the anatomic conditions in the outflow tract of the right ventricle and in the pulmonary orifice. Interpretation of the results is facilitated if the contrast medium is injected directly [illegible]

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[illegible] stressed particularly by Jonsson, Broden and Karnell (369, 371).

We have performed angiocardiography in 54 cases of valvular pulmonary stenosis. Only true frontal and lateral projections were used.

The pathologicoanatomic basis of valvular pulmonary stenosis consists of a conical or domed membrane formed by the fused cusps and provided with a central or slightly eccentric orifice (Fig. 184). In every case the stenosis could be visualized on the contrast examination. It is then seen as a membranous dome, the orifice being outlined by the stream of contrast medium which resembles a jet (Fig. 185). As a rule, the [illegible]

and [illegible]

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The width of the orifice varies in the individual case (Figs. 185, 186 and 187). The membrane formed by the cusps is usually most clearly visualized in the lateral view. In the anteroposterior picture, it is often seen in the semiaxial view, owing to the somewhat dorsal course of the outflow tract. We did not observe the constriction of the annulus described by Currens, Kinney, and White (170) in autopsy cases.

Despite fusion of the cusps, the valve is definitely mobile in the majority of cases, and it is often possible to distinguish the three commissures which, in certain phases of the cardiac cycle, are clearly outlined against the cusps (Fig. 190). The individual cusps are usually most distinct during diastole, but unless the membrane is greatly thickened, they can also be identified during systole (Fig. 190). In the case illustrated in Figure 191, there is distinct asymmetry of the membrane and a local thickening extending to its anterior part. A tumor was assumed to be present, and this was confirmed at operation. It resembled that shown in Figure 192 (p. 199). Such a formation should not be confused with the thickening of the free margins of the valve, which causes a distinct filling defect in diastole (Figs. 193 and 194), but is not particularly conspicuous in systole.

The stenotic valvular membrane is usually thicker than normal valves. This also applies in very mild stenosis with a pressure gradient across the valve of 10 to 15 mm Hg. The degree of thickening is extremely

was below 90 per cent in four cases and below 80 per cent in only one case. In an additional eight cases in which the catheter

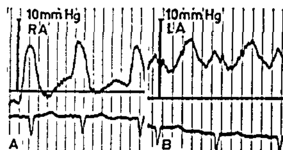


Fig. 182.—Pressure curve from the right (*A*) and left (*B*) atria in a case of valvular pulmonary stenosis. Boy, aged 8 (K.P. 450701). Systolic pressure in ventricle, 230 mm Hg During auricular systole, pressure was somewhat higher in the right atrium, but during the rest of the cardiac cycle it was higher in the left atrium No intracardiac shunt, presumably, only a small patent foramen ovale.

passed into the left atrium, no right to left interatrial shunt was found. In one of them the systolic right ventricular pressure was 230 mm Hg. The interatrial communication was presumably small.

Pressure curves for both atria in this case are recorded in Figure 182. Even if the *a* wave was somewhat higher in the right than in the left atrium, the pressure during the rest of the cardiac cycle was higher in the left atrium When such a patient grows older and right ventricular failure develops, the pressure rises to such an extent that a right to left shunt occurs (591, 713). This presents the characteristic picture of cyanose tardive.

HEMODYNAMICS DURING EXERCISE

Hemodynamics during exercise were studied in only a few children, and only with a low load. Consequently, in order to throw some light on this question, we have selected different types of pulmonary stenosis in adult patients examined at the Department of Clinical Physiology, Karolinska Sjukhuset, and not otherwise included in our series.

Despite the increased resistance in the outflow tract, the right ventricle can—owing to hypertrophy—maintain an ordinary stroke volume both at rest and on exertion In healthy subjects, mechanical systole is shortened during exercise. This shortening can also occur in pulmonary stenosis, but the pressure must then rise still more in order for the stroke volume to remain unaltered. A fairly ordinary stroke volume, both at rest and during exercise, was present in two of the four cases with intact ventricular septum, and the working capacity was normal in both cases (6/55 and 46/57 in Table 4). The two other patients (Cases 56/56 and 25/57) had, on the con-

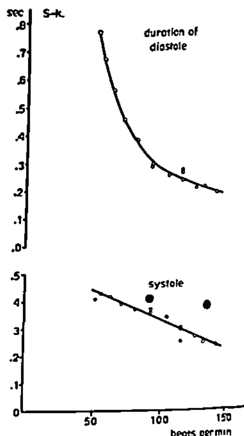


Fig. 183.—Relation between pulse rate and duration of systole at rest and during exercise in severe pulmonary stenosis, Case 56/56 in Table 4 (filled circles), as compared with conditions in healthy subjects (open circles) according to Holmgren (344).

trary, very low stroke volume in the right ventricle even at rest. A considerable shunt from the right to the left atrium was present even at rest, and they exhibited a clini-

cal picture in agreement with that in tetralogy of Fallot. During exercise (Case 25/57) the stroke volume decreased in the right ventricle at the same time that it increased in the left ventricle; i e, the right to left shunt increased, so that the arterial oxygen saturation fell from 83 to 40 per cent. The mechanical systole in the right ventricle could be determined in only one of the cases (56/56). The working capacity was so low that the patient was able to work only for two and one-half minutes with a very low load. The pulse rate then rose to 136 beats per minute, and the arterial oxygen saturation fell from 69 to 54 per cent The stroke volume at work could not be determined. It was, however, evident that the right ventricle was unable to raise the pressure and shorten mechanical systole to the normal extent (see Fig 183). As a result, the shortening of diastole at a high pulse frequency was so considerable that diastolic filling of the ventricle decreased.

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Valvular Stenosis

From the surgical point of view, it is desirable for the angiocardiographic examination to be aimed mainly at elucidating the anatomic conditions in the outflow tract of the right ventricle and in the pulmonary orifice. Interpretation of the results is facilitated if the contrast medium is injected directly into the right ventricle, since this prevents overlapping of the other chambers of the heart on the picture. These considerations have been stressed particularly by Jönsson, Brodén and Karnell (369, 371).

We have performed angiocardiography in 54 cases of valvular pulmonary stenosis Only true frontal and lateral projections were used.

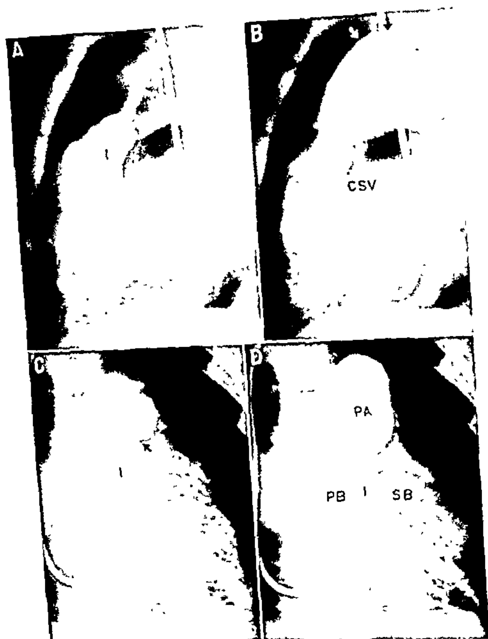
The pathologicoanatomic basis of valvular pulmonary stenosis consists of a conical or domed membrane formed by the fused cusps and provided with a central or slightly eccentric orifice (Fig 181). In every case the stenosis could be visualized on the contrast examination. It is then seen as a membranous dome, the orifice being outlined by the stream of contrast medium which resembles a jet (Fig. 185) As a rule, the orifice is situated centrally (Figs. 185 and 186). Examples of its eccentric localization are shown in Figures 187 and 188. The width of the orifice varies in the individual case (Figs. 185, 186 and 187). The membrane formed by the cusps is usually most clearly visualized in the lateral view. In the anteroposterior picture, it is often seen in the semiaxial view, owing to the somewhat dorsal course of the outflow tract. We did not observe the constriction of the annulus described by Currens, Kinney, and White (170) in autopsy cases.

Despite fusion of the cusps, the valve is definitely mobile in the majority of cases, and it is often possible to distinguish the three commissures which, in certain phases of the cardiac cycle, are clearly outlined against the cusps (Fig. 190). The individual cusps are usually most distinct during diastole, but unless the membrane is greatly thickened, they can also be identified during systole (Fig 190). In the case illustrated in Figure 191, there is distinct asymmetry of the membrane and a local thickening extending to its anterior part. A tumor was assumed to be present, and this was confirmed at operation. It resembled that shown in Figure 192 (p. 199). Such a formation should not be confused with the thickening of the free margins of the valve, which causes a distinct filling defect in diastole (Figs 193 and 194), but is not particularly conspicuous in systole.

The stenotic valvular membrane is usually thicker than normal valves. This also applies in very mild stenosis with a pressure gradient across the valve of 10 to 15 mm Hg The degree of thickening is extremely



Fig. 184.—Valvular pulmonary stenosis, autopsy specimen. Boy, aged 2. *A–B*, marked hyper trophy of trabecular system, ventricular wall and crista supraventricularis, at the ventricula apex, fusion of papillary muscles and of papillary muscles and trabeculae *C*, the cusps, fuse into a dome, seen from the pulmonary artery, in center of the dome, an orifice about 1 mm in di ameter. *CSV*, crista supraventricularis, *PA*, pulmonary artery, *PM*, papillary muscles, *SB*, septa band, *TV*, tricuspid valve.



[illegible]
Cent [illegible]
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to th [illegible]
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the [illegible]
thickness *D*, the contracted parietal and septal bands compress the lower part of the infundibulum The crista supraventricularis bulges in from behind like a powerful muscular ridge *CSV*, crista supraventricularis, *I*, infundibulum, *PA*, pulmonary artery; *PB*, parietal band, *SB*, septal band



Fig. 184.—Valvular pulmonary stenosis, autopsy specimen. Boy, aged 2. *A–B*, marked hypertrophy of trabecular system, ventricular wall and crista supraventricularis; at the ventricular apex, *fusion* of papillary muscles and of papillary muscles and trabeculae. *C*, the cusps, fused [illegible] r of the dome, an orifice about 1 mm in di- [illegible] artery, *PM*, papillary muscles, *SB*, septal [illegible]

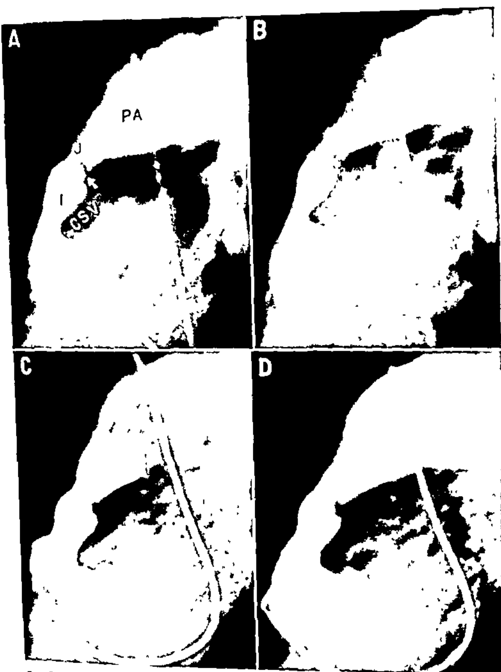


Fig 187 —Valvular pulmonary stenosis Man, aged 45 (B H 080490) Slightly ecc... orifice, about 5 mm ... monary artery ... siderably into ... valvulotomy ... systole, B and ... contrast medium now spurts throug... ably because of decreased hypertrop... PA, pulmonary artery

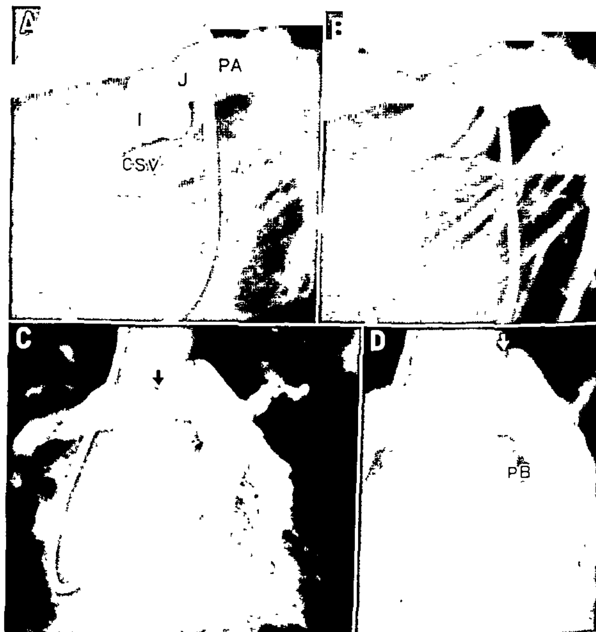


Fig. 186.—Valvular pulmonary stenosis. Girl, aged 10 (A.-L F. 420206). Central orifice, about 10 mm in diameter (*A*) The jet of contrast medium flows centrally in the undilated pulmonary artery. *A*, *C* and *D*, systole, *B*, diastole Cusps inverted, sinuses of Valsalva somewhat irregular, slight thickening of the cusps (*A–B*) Owing to the dorsal course of the infundibulum and pulmonary artery, the valve is difficult to identify in frontal view *C–D*, during systole, marked compression by the parietal band Arrow in *C* indicates narrowing of the right branch of the pulmonary artery (verified at operation) A narrow patent ductus arteriosus causes local dilution of contrast medium in the superior part of the pulmonary artery (arrow in *D*) *CSV*, crista supraventricularis, *I*, infundibulum, *J*, jet, *PA*, pulmonary artery, *PB*, parietal band

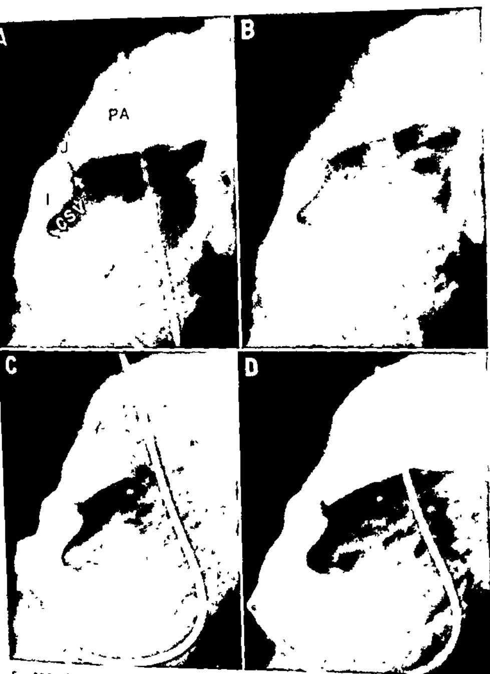


Fig 187.—Valvular pulmonary stenosis Man, aged 45 (B H. 080420). Slightly eccentric
orifice, about 5 mm in diameter The jet flows alo[illegible]
monary artery is seen in A and B Hypertrophi[illegible]
nderably into the lumen Arrow in A marks sit[illegible]
valvulotomy A and C, systole, B and D, diastole [illegible]
contrast medium now spurts through the dilated
ably because of decreased hypertrophy CSV, cri[illegible]
PA, pulmonary artery



Fig. 188.—Valvular st [illegible] upper
arrow in *C* point to the v [illegible]
orifice, about 5 mm in dia [illegible]
(*A*) During injection, a [illegible]
spreading toward the pulmonary valves (lower arrow in *C*) *CSV*, crista supraventricularis, *I*, infundibulum, *PA*, pulmonary artery


A
PA
J
I
CSV
PB
B
C
D
LA
LV
PM

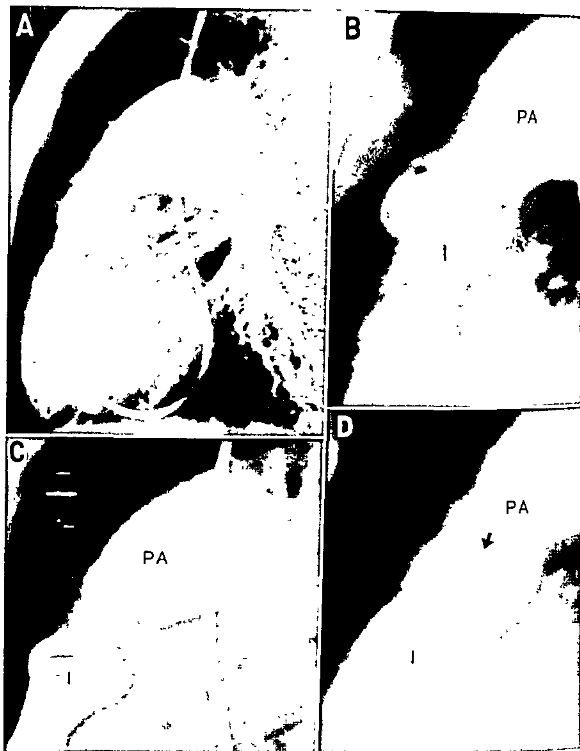


Fig. 190.—Valvular pulmonary stenosis Woman, aged 30 (S A 231205) Central orifice, 12 mm in diameter C, jet directed toward anterior wall of pulmonary artery Main trunk of pulmonary artery dilated distally and anteriorly During systole, marked narrowing of sinuses of Val- [illegible] slit-like space, the three commissures and the cusps are visi- [illegible] rgins cause a small, central defect in the contrast medium [illegible] pulmonary artery



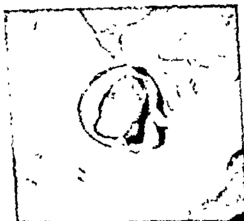


Fig 192 —Autopsy specimen seen from the aorta In the aortic orifice, a fibrous tumor or nating in the noncoronary valve, *it resembles the formation* shown in Figure 191.

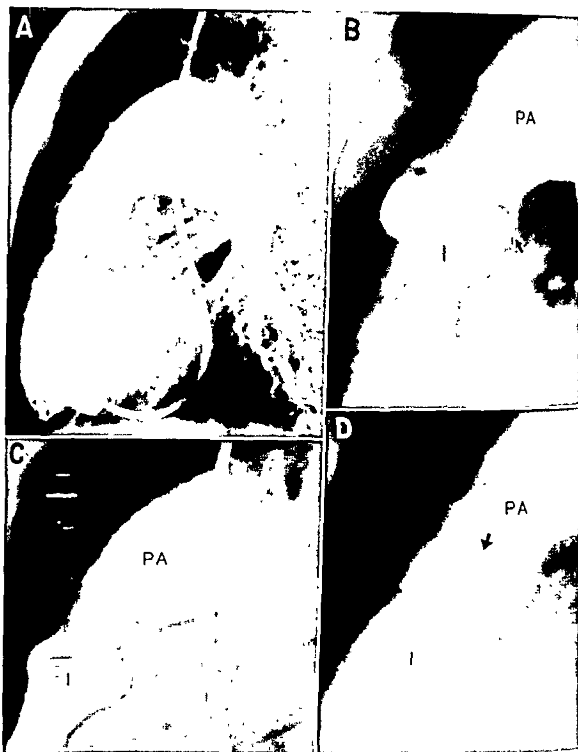


Fig. 190.—Valvular pulmonary stenosis Woman, aged 30 (S A 231205) Central orifice, 1 ... pulm ... Va ... vis ... dur

variable. When the cusps are greatly thickened, an observation which we made in several cases, their mobility is definitely decreased and the membrane is rigid (Figs. 195 and 196). In some of these cases, the valve has a distinctly irregular shape in diastole (Figs 197, 198, *A* and *B*). The [illegible]

direction of the outflow tract (Fig 200).

It can often be demonstrated that the wall of the right ventricle is thickened because of hypertrophy (Fig 201). The outline of the crista supraventricularis, including its septal and parietal bands, is accentuated (Fig 189) and the papillary muscles are prominent (Fig 202). The hypertrophy also involves the infundibulum, this is most evident during *systole* (Figs. 185-189). *In severe stenosis with hypertrophy of the right ventricle*, the septum deviates toward the left ventricle (Fig 197)

During systole, the contraction is *strongly* marked in the ostium *infundibuli*, corresponding to the crista supraventricularis and its bands (Figs 179, 187, 189, 201, 204). If the exposures are made during this phase only, an infundibular stenosis may be simulated Although *the bands* are often hypertrophied, *their* course is normal in valvular stenosis, in contrast to the conditions in infundibular stenosis We did *not* see stenosis of the infundibulum *due to the* hypertrophy in any of our cases.

Interpretation of the findings is greatly facilitated by the rapid succession of the exposures, at least six being made per sec (Figs 189 and 201). However, in several of our cases, despite this frequency various partly random *factors* caused only one or two exposures to be made during diastole, whereas all the *other pictures* were exposed during *systole* or protodiastole Conversely, the majority *of* exposures may be made by chance during diastole (Fig 205).

The main trunk of *the pulmonary* artery is usually dilated, but it may be of normal width, as can be inferred from Figures 188 and 189 This was the rule in children under 4 years of age. The dilatation is consistently most marked distally, whereas some variability is found in the conditions in the proximal part of the vessel. *Occasionally*, there is no dilatation whatever, or the bulb of the artery may be decreased in width (Figs 187 and 191). In other cases the bulb is extremely wide and the sinuses of Valsalva are then prominent (Figs. 200 and 201). In most cases the dilatation is moderate immediately above the valve. The dilatation is continuous in the ventral and lateral segments of the vessel, whereas the transition between the [illegible]

of the main trunk may be considerable (*Fig* 203) and not infrequently involves both main branches as well (Fig. 222), the *branches centrally and peripherally in the* lungs being narrow. In one case in which there was *also* stenosis at the site of origin *of the main* branches, these vessels were greatly dilated, whereas the main trunk *was of ordinary* width (Fig. 139). In rare cases *the central vessels* may also be dilated *but exhibit a* rapid reduction in caliber. Like [illegible]

[illegible] of post-stenotic dilatation. Farthest distally in the *main trunk*, there is often a distinct but [illegible]

During injection of contrast medium, a series of *extrasystoles* often occurs, particularly *when the injection* is made rapidly. *The* essential cause *of this* disturbance in rhythm is *probably the* movements of the tip of the catheter *against* the wall of the ventricle during injection and the contact of the strongly hypertonic contrast *medium with the* endocardium The increase of fluid content of the ventricle due to the contrast medium is, on *the* contrary, presumably of only minor *importance* in the causation of this *arrhythmia. In all* probability, the flow of *medium* into the *ventricle is* immediately compensated by a decreased *inflow* from the atrium. During this [illegible]

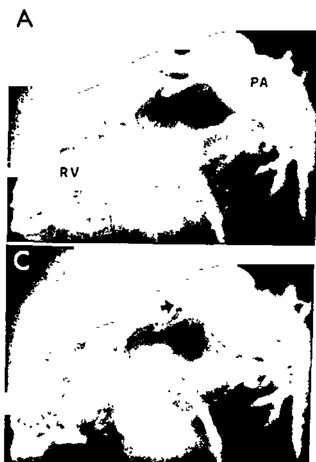




Fig. 193.—Valvular pulmonary stenosis. Girl, aged 1 (B.K. 541216). No poststenotic dilatation of pulmonary artery. The valvular part of the main trunk (bulb of pulmonary artery) is narrow. The thin jet of contrast medium flows centrally in the vessel (arrow in *C*). The cusps are fused into a dome. The margins of the valve are distinct (arrows in *B*). *PA*, pulmonary artery, *RV*, right ventricle.



Fig 194 —Valvular pulmonary stenosis Girl, aged 4 (A-B E 520218) A fairly large, round soft-tissue formation (between arrows in *A*) is visible on the margins of the cusps, it probably represents the thickened free margins of the valve. Slight stenosis (between arrows in *B*) is also seen at the origin of the right main branch

variable When the cusps are greatly thickened, an observation which we made in several cases, their mobility is definitely decreased and the membrane is rigid (Figs. 195 and 196) In some of these cases, the valve has a distinctly irregular shape in diastole (Figs. 197, 198, A and B). The site of greatest thickening may be the margin at the orifice (Fig 199). The valvular plane may be oblique in relation to the main direction of the outflow tract (Fig 200).

It can often be demonstrated that the wall of the right ventricle is thickened because of hypertrophy (Fig. 201). The outline of the crista supraventricularis, including its septal and parietal bands, is accentuated (Fig. 189) and the papillary muscles are prominent (Fig. 202). The hypertrophy also involves the infundibulum, this is most evident during systole (Figs 185-189). In severe stenosis with hypertrophy of the right ventricle, the septum deviates toward the left ventricle (Fig 197)

During systole, the contraction is strongly marked in the ostium infundibuli, corresponding to the crista supraventricularis and its bands (Figs. 179, 187, 189, 201, 204). If the exposures are made during this phase only, an infundibular stenosis may be simulated. Although the bands are often hypertrophied, their course is normal in valvular stenosis, in contrast to the conditions in infundibular stenosis. We did not see stenosis of the infundibulum due to the hypertrophy in any of our cases

Interpretation of the findings is greatly facilitated by the rapid succession of the exposures, at least six being made per sec (Figs. 189 and 201). However, in several of our cases, despite this frequency various partly random factors caused only one or two exposures to be made during diastole, whereas all the other pictures were exposed during systole or protodiastole. Conversely, the majority of exposures may be made by chance during diastole (Fig 205).

The main trunk of the pulmonary artery is usually dilated, but it may be of normal width, as can be inferred from Figures 186 and 189 This was the rule in children under 4 years of age. The dilatation is consistently most marked distally, whereas some variability is found in the conditions in the proximal part of the vessel. Occasionally, there is no dilatation whatever, or the bulb of the artery may be decreased in width (Figs. 187 and 194). In other cases the bulb is extremely wide and the sinuses of Valsalva are then prominent (Figs. 200 and 201). In most cases the dilatation is moderate immediately above the valve. The dilatation is continuous in the ventral and lateral segments of the vessel, whereas the transition between the dilated distal part and the bulb often is distinctly marked in the dorsal segment of the vessel (Fig 190). Dilatation of the main trunk may be considerable (Fig 203) and not infrequently involves both main branches as well (Fig. 222), the branches centrally and peripherally in the lungs being narrow. In one case in which there was also stenosis at the site of origin of the main branches, these vessels were greatly dilated, whereas the main trunk was of ordinary width (Fig 139). In rare cases the central vessels may also be dilated but exhibit a rapid reduction in caliber Like other observers (660), we have been unable to find any direct correlation between the degree of stenosis and the degree of poststenotic dilatation Farthest distally in the main trunk, there is often a distinct but hemodynamically unimportant constriction of the anterior wall of the vessel, caused by the pericardial [illegible]

Dur [illegible] series [illegible] larly [illegible] the injection is made rapidly. The essential cause of this disturbance in rhythm is probably the movements of the tip of the catheter against the wall of the ventricle during injection and the [illegible] of the [illegible] wi [illegible] co [illegible] m[illegible] [illegible] contrary, presumably of only minor importance in the caus[illegible] of this [illegible]



Fig. 195.—Severe valvular pulmonary stenosis and atrial septal defect with large right to left interatrial shunt. Girl, aged 10 (B H 470514) cf Figure 144 Greatly impeded flow



Fig. 195 (*cont*)

through orifice only abou
contrast *medium*, directec
pericardial reflection (arr
tion either into the right
when the flo

ostium (E-F) F
(arrow points to one
jet I, infundibulum



Fig. 195.—Severe valvular pulmonary stenosis and atrial septal defect with large right to left interatrial shunt Girl, aged 10 (B H 470514) cf Figure 144 Greatly impeded flow

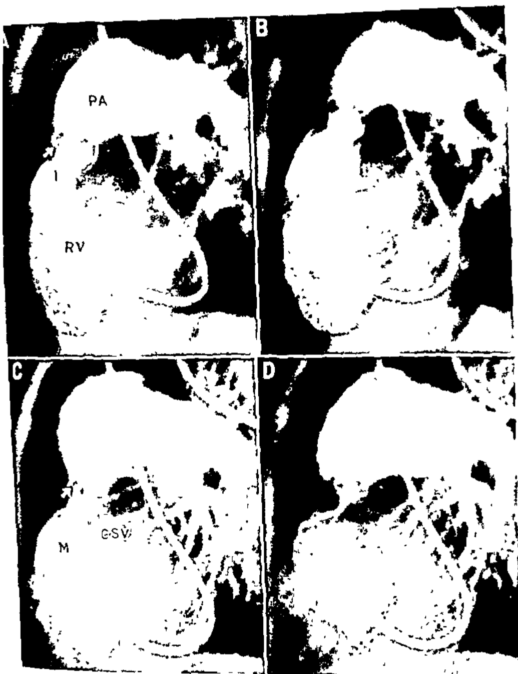


Fig 197.—Valv...
in systole, B

thoracic wal
crista su
downwai



Fig. 196.—Valvular pulmonary *stenosis* Girl, aged 4 (A-C B 511215) Cusps, thick, fused into a dome. Small central *orifice*, *poststenotic* dilatation of main trunk of pulmonary artery Postoperatively (C) good outflow, *decrease* in poststenotic dilatation *CSV*, crista supraventricularis, *I*, infundibulum, *PA*, pulmonary *artery*; *PM*, papillary muscle

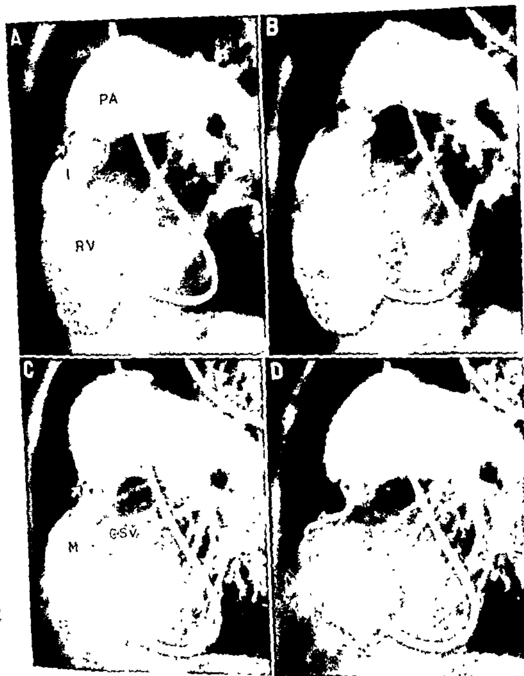

A
PA
I
RV
B
C
M
CSV
D



Fig. 196.—Valvular pulmonary stenosis Girl, aged 4 (A-C.B. 511215). Cusps, thick, fused into a dome Small central orifice; poststenotic dilatation of main trunk of pulmonary artery. Postoperatively (C) good outflow, decrease in poststenotic dilatation CSV, crista supraventricularis, *I*, infundibulum, *PA*, pulmonary artery, *PM*, papillary muscle.



Fig 198a.—Valvular pulmonary stenosis Boy, aged 8 (K P. 450701) Cusps are thick and rigid and undergo only inappreciable changes during the cardiac cycle *A* and *C*, systole, *B*, [illegible]

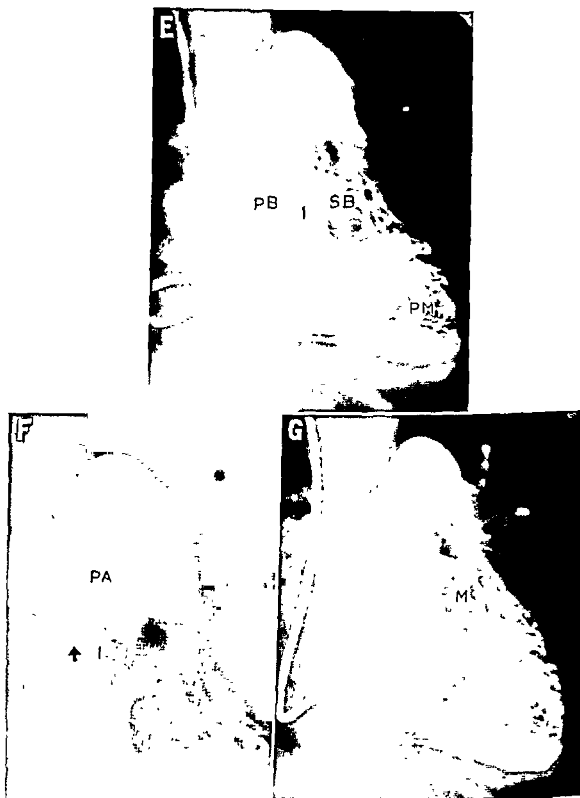


Fig. 197 (*cont*)

traction of the crista and of both bands on the infundibulum is most distinct in comparisons between *E* and *G* and between *B* and *C*. No retraction of anterior wall, but lifting of apex and caudal part of the ventricle, corresponding to the attachment of the parietal and septal bands. *CSV*, crista supraventricularis, *I*, infundibulum, *M*, muscular ridge, *PA*, pulmonary artery; *PB*, parietal band, *PM*, papillary muscle, *RV*, right ventricle. *SB*, septal band



Fig 200 —Valvular pulmonary stenosis. Girl, aged 5 (B N. 480527). Flattened dome, valvular plane oblique in both systole (*A*) and diastole (*B*), no narrowing of sinuses of Valsalva, hypertrophied papillary muscles distinct in both pictures *I*, infundibulum, *PA*, pulmonary artery, *PM*, papillary muscles.

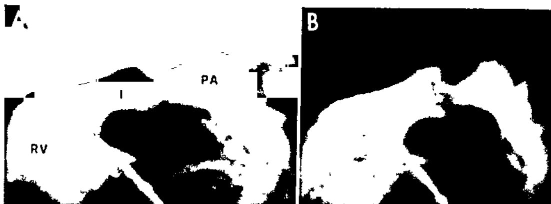


Fig. 198b.—Valvular pulmonary stenosis. Boy, aged 2 (H.L. 540305). Irregular, thick, and rigid cusps. The part of the main trunk close to the valve (bulb of pulmonary artery) is very narrow; no poststenotic dilatation of pulmonary artery. *A*, systole, *B*, diastole. *I*, infundibulum; *PA*, pulmonary artery; *RV*, right ventricle

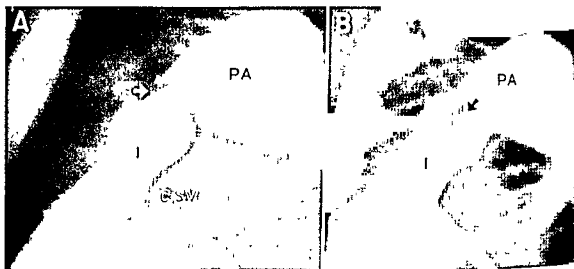


Fig. 199.—Valvular pulmonary stenosis Boy, aged 2 (K W. 510629) *A*, systole, *B*, diastole Central orifice, about 3 mm in diameter, cusps mobile Arrow in *A* points to valvular plane, arrow in B, to defect in contrast medium formed by the valvular margins *CSV*, crista supraventricularis, *I*, infundibulum, *PA*, pulmonary artery

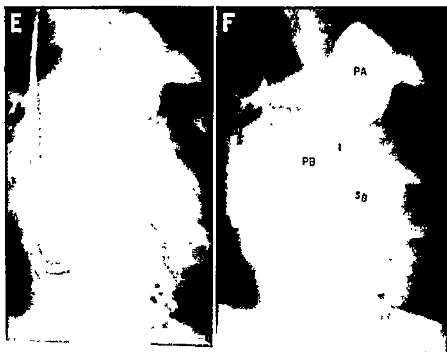
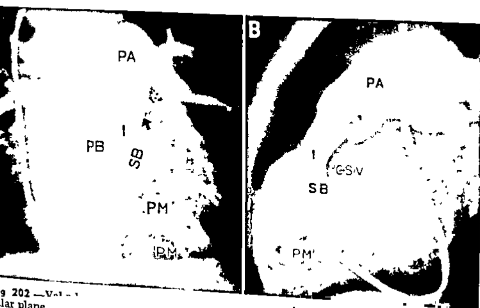


Fig. 201 (*cont.*)



...g 202 —V...
...lar plane
...lary musc
... PA, pulm

... band, PM, papillary muscles, SB, septal band.

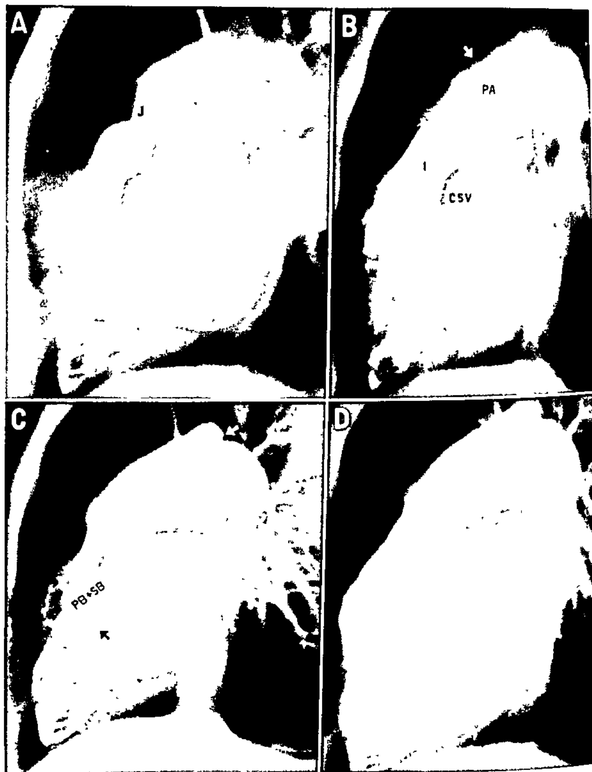


Fig. 201.—Valvular pulmonary stenosis Girl, aged 6 (B L 440127). Through the central orifice in the dome, a 5 mm wide jet of contrast medium spurts toward the anterior wall of the pulmonary artery, which bulges at the site of impact (arrow, B) During the injection, powerful
·w, bands partly over-
arrow marks caudal
he ventricular wall is
ulmonary artery, PB,

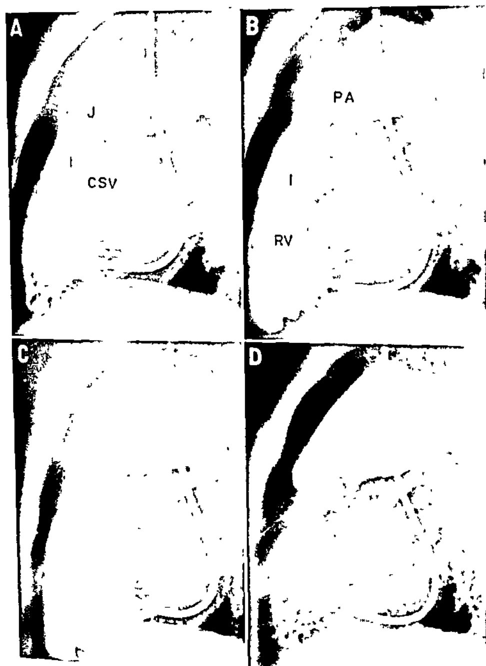


Fig 204.—Valvular pulmonary stenosis Boy, aged 11 (L.W. 421111). Central orifice. The jet is directed toward anterior wall of pulmonary artery and accompanies it as far as ... dual reflection Here, it turns caudally (A). ...

... right ventricle ... I, infundibulum. J, jet,

strong contractions are often induced in the right ventricle, particularly at the level of the ostium infundibuli. As a result, the infundibulum is almost shut off from the sinus region of the ventricle and assumes the appearance of a balloon (Fig. 204). After the contrast medium has been deposited in the ventricle, the rhythm of the heart usually returns to normal.

Thus, some of the findings at angiocardiography are influenced by the technique. This has little effect on the interpretation of the purely anatomic conditions, provided exposures are obtained from the different phases of the cardiac cycle Conclusions regarding the hemodynamic conditions should, on the other hand, be drawn with caution. Certain phenomena nevertheless seem to be well illustrated by angiocardiography.

The blood passes through the stenotic pulmonary orifice in the form of a jet. It is usually directed toward the anterior wall of the pulmonary artery, in a section a few centimeters above the valvular plane, where a shallow bulging of the wall marks the impact of the jet (Figs. 201, 204, 206, and 207). In several cases this local bulge was situated high up in the pulmonary artery (Figs. 208, 209, and 210). Although the patients are in the supine position during angiocardiography, which, in view of the weight of the contrast medium, could be expected to result in initial filling of the posterior segment of the pulmonary artery, it is evident that the early opacification takes place in its anterior and cephalic segments only. Distinct turbulence is often seen during the spreading of the contrast medium (Figs. 185, 201, 204–206). The posterior part of the vessel becomes more completely filled only at a later stage. Occa-

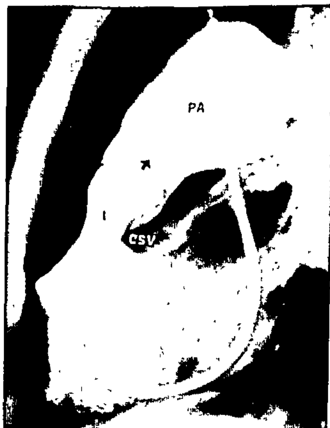


Fig. 203.—Valvular pulmonary stenosis Girl, aged 10 (E I 440429) Central orifice (arrow), about 5 mm in diameter, great dilatation of pulmonary artery *CSV*, crista supraventricularis, *I*, infundibulum, *PA*, pulmonary artery

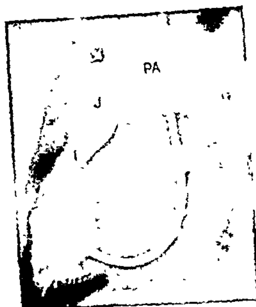


Fig. 206.—Valvular pulmonary stenosis Boy, aged 10 (G S. 431126) The jet is directed toward the anterior wall of the pulmonary artery, arrow marks site of impact. Dome is flat, with a central orifice, about 5 mm in diameter, eddies in the artery I, infundibulum, J, jet, PA, pulmonary artery

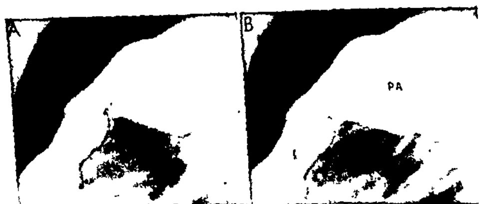


Fig 207.—Valvular pulmonary stenosis Boy, aged 10 (A P-P. 440403) The 5–6 mm wide jet impinges on the anterior wall of the main trunk and causes it to bulge outward *PA*, pulmonary artery, *I*, infundibulum

sionally, the course of the jet in the artery is axial Local dilatation of the vessel may then be lacking (Fig 186) or it may be situated more superiorly and be less distinct (Fig. 185) With a greatly reduced cardiac output e g. in heart failure, we observed in one case of severe stenosis that the jet was damped directly above the valvular plane (Fig 211)

The following picture of the function is obtained in the valvular region and the right ventricle During the protosystolic phase, there is protrusion of the fused cusps —the dome—into the pulmonary artery (Fig 212) Figley (253) has pointed out that this

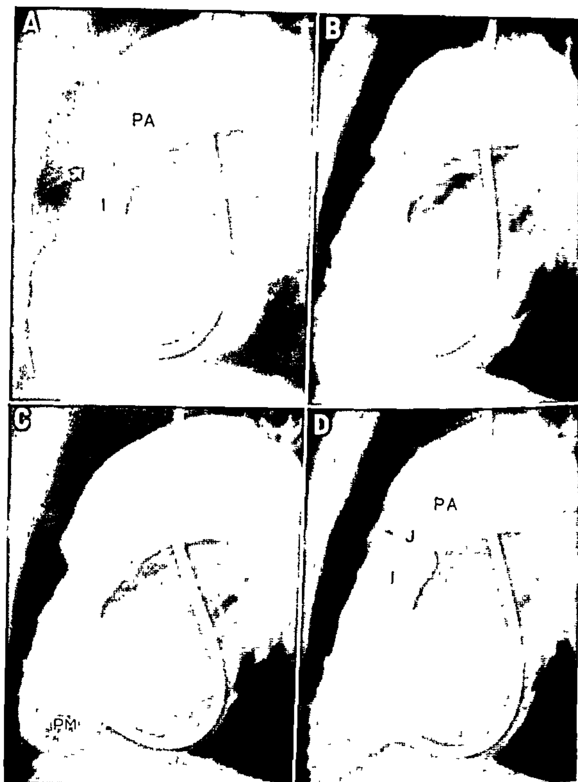


Fig. 205.—Valvular pulmonary stenosis Girl, aged 7 (K D. 460418). Centrally in the fairly flat dome, there is an orifice about 5 mm in diameter, through which the contrast medium pours without actually forming a jet (*D*). Turbulence in the pulmonary artery visible in all views, inappreciable dilatation of pulmonary artery This is an example of the necessity of a rapid succession of exposures. In this case, despite 6 exposures/second, only one exposure was obtained in systole to five or six in diastole The appearance during systole definitely established the diagnosis. *I*, infundibulum, *J*, jet, *PA*, pulmonary artery, *PM*, papillary muscle

cases. The membrane is seen to extend farther into the vessel than do normal valves. It is expanded and may be presumed to convey an impact to the blood in the trunk of the pulmonary artery, which is slightly dilated In some cases, the dome compresses the sinuses of Valsalva into a narrow, slit-shaped space (Fig 185). The low pressure that occurs beside the jet may possibly contribute to deficient expansion of the sinuses of Valsalva, but it may be noted that their size is not correlated with the degree of stenosis The initial displacement of because we endeavored in every case to deposit the contrast medium in the right ventricle. In only one case, in which the tip of the catheter caused bursts of extrasystoles on entering the ventricle, was the injection made into the right atrium In eight cases, the tip recoiled into the atrium at the end of the injection, so that it was made partly into this chamber. A distinct passage of the contrast medium into the left atrium, because of a right to left shunt, was seen in only three of these cases Experience from intravenous angiocardiography shows



Fig. 209 —Valvular pulmonary stenosis and atrial septal defect. Boy, aged 14 (C-A H 410928) A powerful jet spurts through the 5 mm wide orifice toward the anterior wall of the main trunk, which bulges outward. Considerable turbulence in pulmonary artery.

the membrane is completed slightly after the S wave in the electrocardiogram. In the subsequent phase, corresponding to the phase of ejection, a caudal displacement takes place in the valvular plane and there is further dilatation of the pulmonary artery.

During diastole, the valvular membrane is inverted and bulges toward the lumen of the ventricle. During this phase, the appearance of the fused cusps differs not at all, or only slightly, from that of normal valves When the cusps are rigid, no inversion of the valvular membrane occurs and it has an irregular appearance (Figs 196 and 198)

From the angiocardiographic point of view, we have only limited experience of the anatomy and hemodynamics of the right atrium in pulmonary stenosis. This is that an initial, massive filling of the left side through the right atrium may cause considerable difficulty in judging of the anatomic conditions in both the pulmonary orifice and the aortic root. In the patients in question, only the latter part of *the injection* was made into the atrium, and the contrast filling of the left side was delayed to the corresponding extent. Over-riding of the aorta could therefore be ruled out, particularly since the aortic root was at the normal site. In all of the eight cases in which the right atrium was visualized on angiocardiography, it was enlarged. The enlargement was considerable in two of these cases and the auricular [illegible]

Leaki[illegible]
right ve[illegible]
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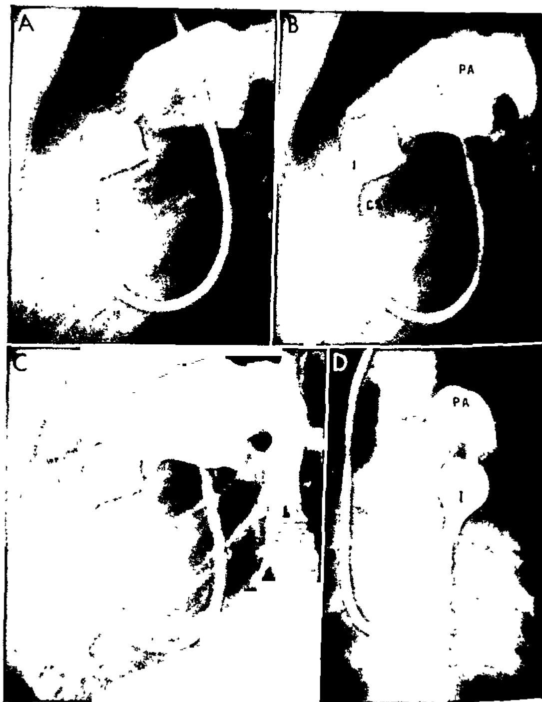


Fig. 208.—Valvular pulmonary stenosis Woman, aged 20 (M.S. 350519) Orifice 3 mm wide Great turbulence and slow mixing in main trunk of pulmonary artery The jet impinges on the wall of the artery at some distance from the orifice *A–C*, lateral view, *D*, frontal view. *CSV*, crista supraventricularis, *I*, infundibulum, *PA*, pulmonary artery

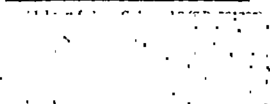












ment of the septal band (continued)



Fig. 210.—Valvular pulmonary stenosis Man, aged 23 (I J 330926) The powerful jet (*J*) spurts far up into the pulmonary artery, which is greatly dilated in front of the pericardial reflection (arrow in *B*). This segment has a recess-like appearance The dilatation does not involve the branches of the pulmonary artery Discrepancy in shape of the sinuses of Valsalva



Fig 213 —Supravalvular pulmonary stenosis Boy, aged 9 (L W 440827) At level of the ostium of the pulmonary bulb, a transverse, thin, membranous stenosis Pulmonary cusps, about 1 5 cm caudal to it, are intact, they open and close normally A, systole, B, diastole. In B and C, lower arrows point to valvular plane, upper arrows to the membranous constriction Inappreciable poststenotic dilatation of pulmonary artery D, late diastole The large papillary muscles are seen clearly in E, the posterior is marked PM, that situated anteriorly is immediately above it.

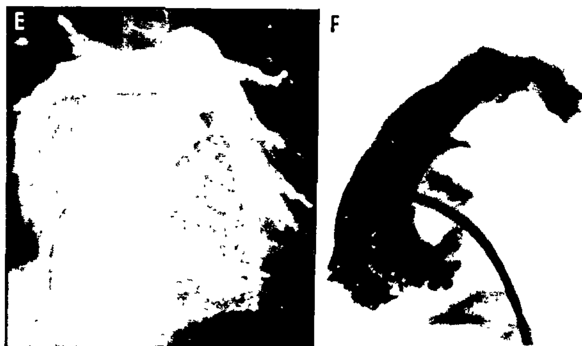


Fig. 211 (*cont*)

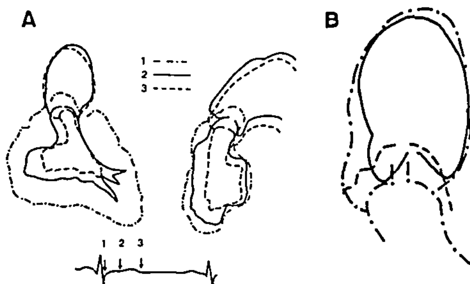


Fig. 212.—Valvular pulmonary stenosis. Boy, aged 6 (T J. 471008), see Figure 185 *A*, diagram of mode of contraction in the right ventricle, caudal part is lifted, pulmonary artery with valvular plane is drawn caudally, and infundibulum is compressed from both the posterior direction and the sides *B*, variation in position and shape of the pulmonary artery and the valvular plane during the cardiac cycle Solid line, late diastole, broken line, early systole, broken-dotted ... pulmonary artery ... d cau



Fig 213 —Supravalvular pulmonary stenosis Boy, aged 9 (L W 440827). At level of the ostium of the pulmonary bulb, a transverse, thin, membranous stenosis Pulmonary cusps, about 1 5 cm caudal to it, are intact, they open and close normally *A*, systole, *B*, diastole In *B* and *C*, lower arrows point to valvular plane, upper arrows to the membranous constriction. Inappreciable poststenotic dilatation of pulmonary artery D, late diastole The large papillary muscles are seen clearly in *E*, the posterior is marked *PM*, that situated anteriorly is immediately above it.

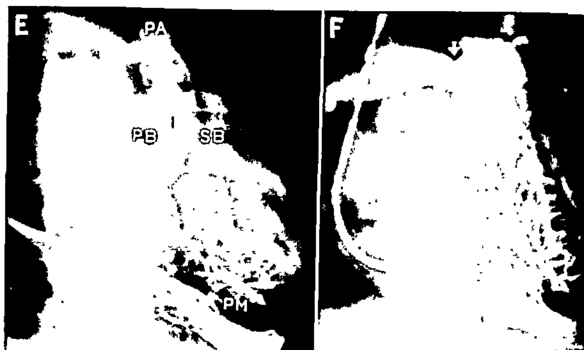


Fig. 213 (*cont.*)
Slight constriction of the right branch of the pulmonary artery (arrow in *F*), verified at operation. *CSV*, crista supraventricularis, *I*, infundibulum, *PA*, pulmonary artery, *PB*, parietal band, *PM*, papillary muscle, *SB*, septal band

by the catheter and the resulting leakage of the orifice (Fig 201). Consequently, this phenomenon should not be attributed without reservation to tricuspid incompetence.

In many cases, advantage was taken of the quantity of contrast medium that had passed through the pulmonary circulation for study of the left atrium, left ventricle, and aorta In every case the left side exhibited a normal configuration. In a few cases, however, the left atrium was small and the aorta narrow Figure 147 shows how, owing to its position, the undilated left atrium causes displacement of the esophagus and a bulge in it Similar observations were made in five cases. In such cases, backward displacement of the left atrium on conventional roentgenograms should not be taken as evidence of enlargement

One of the physical signs of right ventricular hypertrophy is a parasternal lift. Its anatomic basis is clearly illustrated by the findings on angiocardiography It can be seen in Figures 197 and 204 how, during systole, the curvature of the right ventricle is accentuated so that it bulges toward the thoracic wall, presumably transmitting an impulse to it which, on palpation, is felt as a lift

Supravalvular Stenosis

Our series includes one case of supravalvular stenosis, it was diagnosed at angiocardiography and confirmed at operation (Fig 213) Above the valve, which is normal as regards appearance and mobility, is seen a septum-like constriction, it appears as a narrow contrast defect in the lumen During the cardiac cycle, the membrane exhibits only inappreciable mobility.

Infundibular Stenosis

Morphologic background —According to Edwards (214), infundibular stenosis is characterized by "localized stenosis of the outflow tract of the right ventricle. The level of the stenosis usually lies several centimeters inferior to the pulmonary orifice,



Fig 214 —Infundibular stenosis with displacement of parietal band (type I). A–C, right ventricle, D, left ventricle With the right part of the crista supraventricularis, the parietal band is

cusp of the tricuspid valve. Owing to the change in position of the parietal band and crista supraventricularis, the ostium infundibuli (arrow in C) is compressed from the right and behind

ventricular septum

thereby converting the right ventricle into two parts." Lev (424) has classified infundibular stenosis into two types. He wrote:

1. There may be a fibrotic band at the line of junction of the conus and the sinus. This produces a stenosis or atresia of the orifice of the conus, and a division of the right ventricle into two. The conus may be enormously dilated above the constriction.

This type is seen in transposition complexes, or associated with a ventricular septal defect without transposition

2. The entire conus may be shrunken in size. An abnormally formed crista meets the anterior wall to produce a shrunken conus with thick walls, constituting a stenotic outlet to the right ventricle The endocardium in this region is usually thickened.

This type of conus is commonly seen in transposition complexes

Lev also pointed out that in most cases of tetralogy of Fallot the parietal portion of the crista (parietal band) is "foreshortened and deviated towards the left over the anterior wall of the right ventricle. It is now no longer in close proximity to the anterior tricuspid leaflet. The septal portion of the crista (septal band) is more prominent than normal."

Our own observations on autopsy specimens, as well as our findings at angiocardiography in infundibular stenosis without over-riding aorta, have led us to infer that the septal portion of the crista supraventricularis may—on the grounds of foreshortening and an abnormal course—also be involved in the stenosis, either alone or in combination with the parietal portion

We have considered it justified to classify infundibular stenosis without over-riding aorta into two main types, on the basis of the morbid anatomy of the infundibular bands. One of the chief advantages of this classification is that it facilitates the interpretation of the angiocardiographic observations

1. In the first type, which is the more common, the right portion of the crista with the *parietal* band runs in an abnormal direction, it courses ventrally toward the left, and its attachment to the anterior wall of the ventricle is farther up toward the base than usual (Fig. 214) This change in posi-

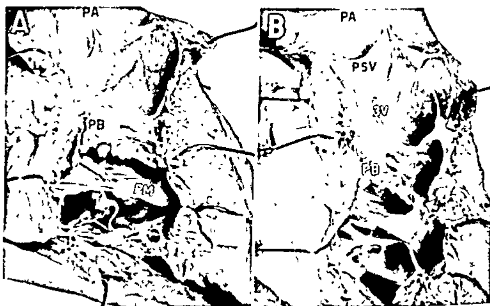


Fig. 215.—Infundibular stenosis (type I). Considerable displacement of parietal band and constriction of ostium infundibuli, which has endocardial thickenings. Third ventricle is wide, considerable hypertrophy of trabeculae and of muscle wall in the sinus region; wall of third ventricle of normal thickness, large pits and channels in left part of ostium infundibuli (arrows in *B*) *PA*, pulmonary artery, *PB*, parietal band, *PM*, anterior papillary muscle, *PSV*, pulmonary semilunar valves, *3V*, third ventricle



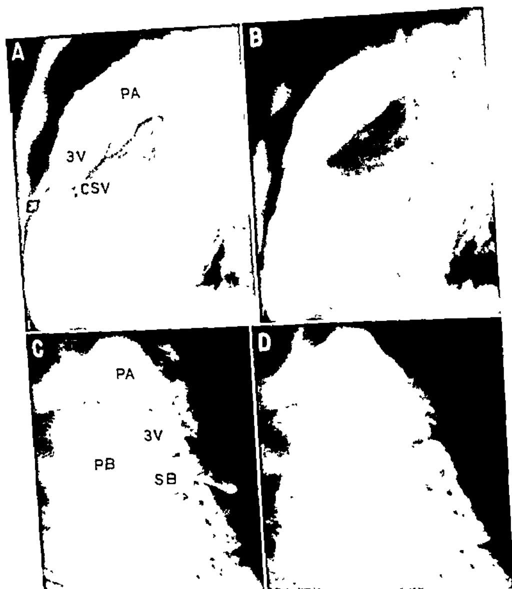
Fig 216 —Infundibular stenosis (type II) and large atrial septal defec... ostium atrioventriculare commune type Boy, age... A–D, right ventricle, E, left atrium, F, right atriu... infundibuli, owing to displacement of the septal... wall caudally, but runs anteriorly and to the right, ... at the level of the crista supraventricularis, it exten... septal defect at site of ostium primum (arrows) ar... tends as far as the ventricular septum (E) ... superior ... crista ... nary a ... tricula ...

tion causes circumscribed constriction of the infundibulum at the ostium from behind, from the right and to some extent *from in front (Figs. 214 and 215)*. Consequently, the degree of stenosis of the infundibulum is essentially dependent on the extent of displacement of the parietal band On embryologic grounds, it is understandable that this type of infundibular stenosis is often associated with a ventricular septal defect. Owing to the displacement of the crista and the parietal band ventrally and to the left, the normal closure of the interventricular foramen is incomplete or does not take place. The size of the septal defect is partly correlated with the degree of displacement. We studied six autopsy specimens of this type.



Fig. 217.—Normal right ventricle Boy, aged 6 (R I. 470521). During diastole, the infundibulum is dilated into a funnel and its width exceeds that of the pulmonary artery (*A* and *D*) During [illegible] into a tube of fairly even thickness, width is then less than that of the [illegible] ormal, open valves and to sinuses of Val- [illegible] *I*, infundibulum, *PA*, pulmonary artery,

2. In the second type, which is presumably not so common, the *septal* band runs ventrally and to the right and joins the anterior wall of the ventricle more superiorly (Figs. 216 and 399b, p 432). The left portion of the crista is displaced ventrally. The infundibulum is thereby compressed from behind and from the left, as well as from in



[illegible]

dibuli Pulmonary valve normal CSV, crista supraventricularis, *PA*, pulmonary artery, *PB*, parietal band, *SB*, septal band, *3V*, third ventricle.

front. The hypertrophied parietal band has a normal course and there is no associated ventricular septal defect. We have not been able to ascertain the incidence of this isolated anomaly in the course of the septal band. Two such cases are present in our autopsy material.

A *combined form*, in which both hypertrophied bands of the crista have an abnormal course, is presumably more common. We found three instances in our autopsy material.

A common feature in all these forms is the *local constriction* of the ostium infundibuli.

Fusion both of the trabeculae below the ostium infundibuli and of the septal band with the papillary muscles has been described, and these conditions are illustrated in Figure 214. Similar anomalies, covered by thickened endocardium and containing small pits and channels, are frequently present at the ostium infundibuli and in the lower part of the third ventricle (Figs. 214 and 216).

ANGIOCARDIOGRAPHY.—As a rule, the stenosis is confined to the part around the ostium infundibuli Transitional cases are found between a tetralogy of Fallot and a ventricular septal defect with a left to right shunt Angiocardiography should therefore be performed by injection of the contrast medium directly into the right ventricle. This will permit visualization of the anatomic conditions in the outflow tract and demonstration of a possible ventricular septal defect and the relation of the aortic root to it Associated valvular stenosis will also be depicted.

The tip of the catheter should, if possible, be placed in the apex or, in any event, below the stenosis If the tip lies in the third ventricle, its anticipated retraction into the sinus part of the ventricle during the injection may not take place. The stenosis will then not be exposed.

For the same reasons as those brought forward in dealing with valvular pulmonary stenosis, it is essential that exposures be made in many phases of the cardiac cycle During systole—particularly in its final stage—difficulty may be encountered at angiocardiography in distinguishing between an infundibular stenosis and a normal infundibulum in a state of contraction. The findings during diastole are to some extent decisive for the interpretation During this phase, the normal infundibulum is dilated into a funnel shape, and its width usually exceeds that of the pulmonary artery (Fig. 217). In stenosis, the ostium infundibuli is never completely dilated During diastole as well, the bands compress the lumen, and the anterior wall of the ventricle is retracted at the level of the insertion of the infundibular bands. The course of the bands is also changed (Figs. 218 and 219).

In our opinion, 10 of the 14 cases referred to this group represent the aforementioned combined form of infundibular stenosis, for both the parietal and the septal portion of the crista exhibit an abnormal course on the angiocardiogram. Four of them represented simple stenosis with an intact ventricular septum. In the other six a ventricular septal defect could be demonstrated; it was situated in the membranous part of the septum. The aorta was not over-riding.

In the remaining four cases, the essential cause of constriction of the ostium infundibuli was a displacement of the septal band, whereas the effect of the parietal band was slight (Fig. 220). The septal band was hypertrophic, it was greatly displaced to the right and shortened. It produced constriction of the lumen of the ostium infundibuli in the posterior and left parts and, to a lesser extent, in the anterior segment The ventricular septum was intact in all these cases. Thus the total number of cases of simple infundibular stenosis, with or without valvular stenosis, in which angiocardiographic examination was made amounts to eight.

Infundibular stenosis caused by a change in the course of the parietal band only was diagnosed in six cases In every case there was an associated ventricular septal defect, and the degree of stenosis was so slight that a left to right shunt was present. These



valve, 3V, third ventricle

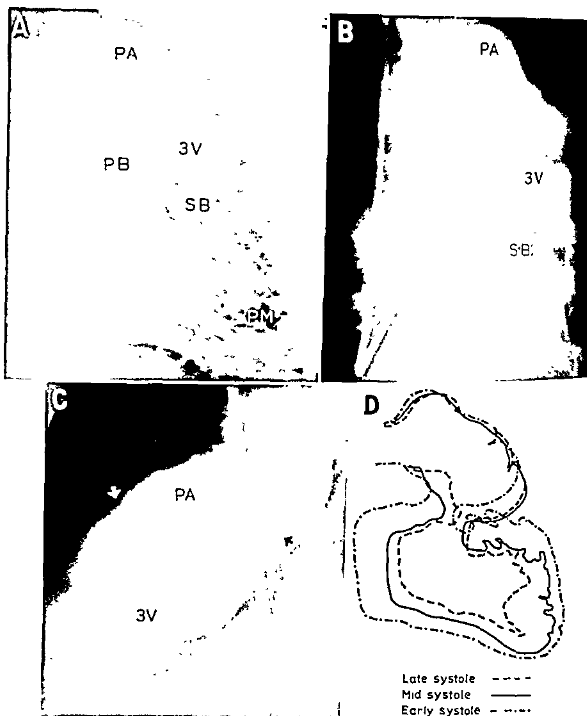
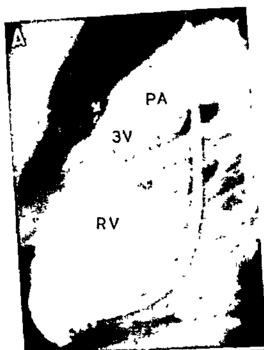
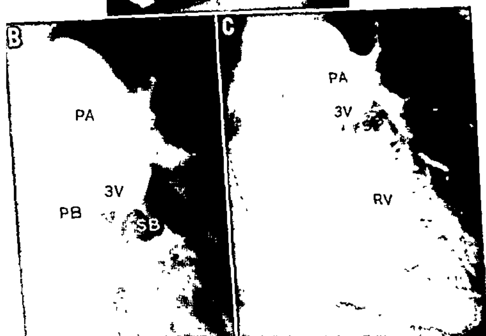


Fig. 220.—Infundibular stenosis Man, aged 28 (K O 240620) Displacement mainly of septal band, but to slight degree of parietal band also Third ventricle large and encircles pulmonary artery (arrows in *C*) In *B*, a tubular bulge in third ventricle overlaps the pulmonary artery (cephalad to *3V*) In *A*, narrow channels are visible at level of septal band. Sinus region is large and extends to left contour of the heart. In *C*, bulge on anterior aspect (below *3V*) represents abnormal insertion of septal band *D*, diagram of mode of contraction of right ventricle During systole, considerable displacement of parietal band caudally and to left, septal band moves in abnormal direction, to left but only inappreciably caudad, incomplete contraction of both sinus region and third ventricle, large residual blood volume *PA*, pulmonary artery, *PB*, parietal band, *PM*, posterior papillary muscle, *SB*, septal band, *3V*, third ventricle

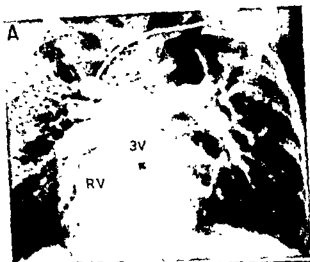




nght ventricle, *SB*, septal band, *3V*, third ventricle.



Fig. 221b.—Severe simple infundibular stenosis. Boy, aged 10 (K D. 470602). The ostium infundibuli and adjacent part of the infundibulum are severely stenosed by the circularly constricting parietal and septal bands of the crista supraventricularis, which are greatly displaced and hypertrophied. The blood leaves the sinus of the right ventricle in a narrow jet, which impinges on the anterosuperior wall of the poststenotically dilated pulmonary artery No stenosis of the pulmonary valve. Ventricular septum intact The right ventricle is dilated and hypertrophied. *PA*, pulmonary artery, *RV*, sinus region of right ventricle Angiocardiograms were made after transventricular infundibulectomy that did not relieve the obstruction Later radical resection was performed by open method. This confirmed that no ventricular septal defect was present.



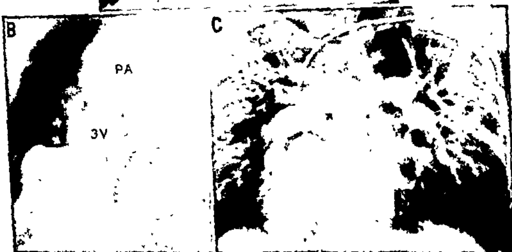


Fig 222 —Infundibular and valvular pulmonary ... 13 (G M 391123) Fairly small third ventricle betw... arrow in C) and infundibular stenosis (lower arrow... compresses the lumen considerably from the right (...

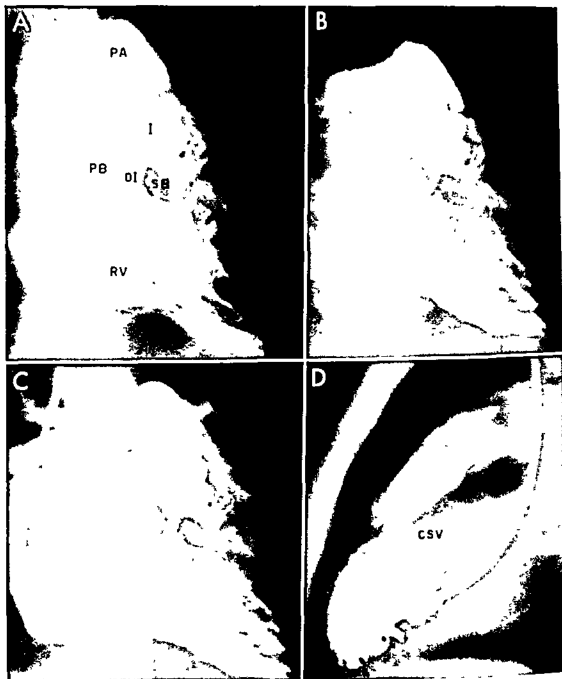


Fig. 223.—Simple infundibular stenosis Girl, aged 7 (M H 470414) Considerable displacement of both parietal (*PB*) and septal band (*SB*) with great constriction of ostium infundibuli (*OI*) both from the sides (*A–C*) and from in front (*D*) Infundibulum of normal width Ventricular septum intact *I*, infundibulum, *PA*, pulmonary artery, *RV*, right ventricle, *CSV*, crista supraventricularis.

cases were therefore classified as ventricular septal defects and are dealt with on page 393.

As may be inferred from Figure 220, D (cf. Fig. 91, p. 87) the abnormal course and insertion of the septal band change the mode of contraction in the infundibulum.

The size of the third ventricle varied considerably It was small in three cases (Fig. 222), and in 12 its width during diastole was the same as that of the normal infundibulum (Figs 218, 219, and 223). In the remaining case, the infundibular chamber was definitely dilated; it encircled the base of the pulmonary artery (Fig 220) and showed small variations in volume In the other cases, the third ventricle varied distinctly in size during the cardiac cycle

Protruding soft tissue formations which contained small cavities in the region of the ostium infundibuli are shown in Figures 219 and 221. They were probably caused by fusion of the trabeculae and possibly by thickened endocardium.

The main trunk of the pulmonary artery was dilated in only three cases (Fig. 221, A); in two of them it was associated with valvular stenosis (Fig 222). In the four other cases in which this combination was present, the pulmonary artery was not dilated.

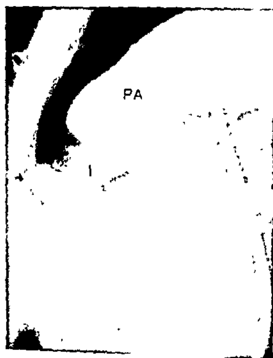


Fig. 224.—Infundibular stenosis. Boy ... valvulotomy) pres... dibular stenosis, ... dilatation remains ... artery.

Figure 224 shows a postoperative aneurysm in the right ventricle. Similar dilatation of the ventricular wall was observed in cases without operation during the actual injection of contrast medium into the ventricle. It is probably produced by the strong, local contractions which spread in waves toward the base of the ventricle (Fig. 100

7 Pulmonary Atresia with Intact Ventricular Septum

PULMONARY atresia generally occurs in association with other malformations, such as ventricular septal defect with over-riding aorta ("pseudotruncus") and tricuspid atresia. As an isolated malformation, it is exceedingly rare. The circulatory conditions are the same as in tricuspid atresia, in so far as the right atrium is drained only through a patent foramen ovale or a possible atrial septal defect. The pulmonary circulation is maintained only through a patent ductus arteriosus or, to the extent to which it has had time to develop, by a collateral circuit. Functionally, this is equivalent to a single ventricle, the right ventricle then being merely a blind cavity. In most cases the right ventricle is small (184, 216, 569), but it may be of normal size or even abnormally large (222, 301). The highly pathologic circulation leads to death in early infancy.

Our series contains two such cases, in one of them the right ventricle was of normal size and in the other it was small. In both cases the wall of the right ventricle was greatly hypertrophied

GIRL, AGED 3 MONTHS (A K 540610) —She was premature, with a birth weight of 2 1 kg She was cyanotic from birth, and heart disease was diagnosed on the first day of life. She gained normally in weight The *cardiac findings* were characterized by a pure second sound and a systolic murmur over the apex The *electrocardiogram* showed marked right ventricular hypertrophy but no pathologic P waves

Roentgenologic examination —The heart was moderately enlarged; enlargement was due chiefly to distention of the right atrium. The anterior border of the right ventricle showed increased curvature, as in hypertrophy. A distinct bulge was present in the left superolateral border of the ventricle (Fig. 225a). The main trunk of the pulmonary artery could not be visualized, and the vascularity of the lungs was sparse. The aorta was judged to be of ordinary width.

Cardiac catheterization —Pressure was raised in the right ventricle (100/8 mm Hg) but normal pressure in the right atrium, with no abnormally high *a* waves (6 mm Hg) The catheter did not pass into the left atrium, and arterial puncture was unsuccessful No left to right shunt was present at the atrial or ventricular level.

Angiocardiography —Contrast medium was injected into the right ventricle (Fig 225b) The infundibulum and sinus region of the ventricle were slightly dilated. The pulmonary artery did not become filled The contrast me[illegible] abnormally long time [illegible]

was incorrectly interpreted by us as an expression of decreased cardiac output, owing to the numerous arrhythmias which occurred during angiocardiography

Autopsy —The patient died two months later, and autopsy showed a right ventricle of normal size with hypertrophy of the wall (Fig 225c) The ventricular septum was intact, and the atrioventricular valves were normal. The pulmonary orifice was atresic, as was the main trunk of the pulmonary artery. The foramen ovale was fairly large

GIRL, AGED 2½ MONTHS (F.H 570608) —She had been cyanotic since birth. She was easily fatigued on feeding, but gained nor-

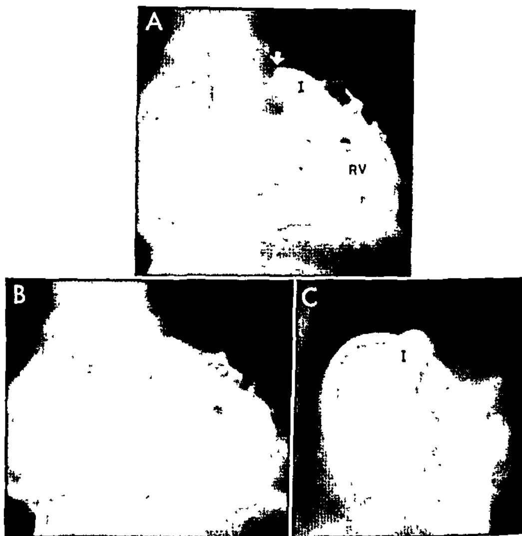


Fig. 225b.—Same case as in Figure 225a. Right ventricle large, with marked hypertrophy of wall muscles Atresia at level of pulmonary orifice (arrow in *A*) Ventricular septum intact Leakage to right atrium through orifice of tricuspid valve. *I*, infundibulum, *RV*, right ventricle

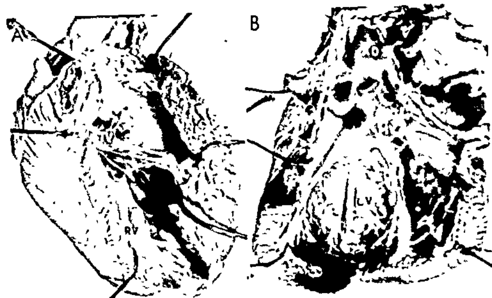


Fig 225c.—Same case as in Figures 225a and 225b Great hypertrophy of muscles of right ventricle Atresia at level of pulmonary orifice (arrow). Right ventricle otherwise of normal shape Ventricular septum intact Atresia of main trunk of pulmonary artery. Its main branches are present but are narrow. Ductus arteriosus patent and fairly wide and opens into left main branch of pulmonary artery *TV*, tricuspid valve, *RV*, right ventricle, *AO*, aorta, *MV*, mitral valve, *LV*, left ventricle

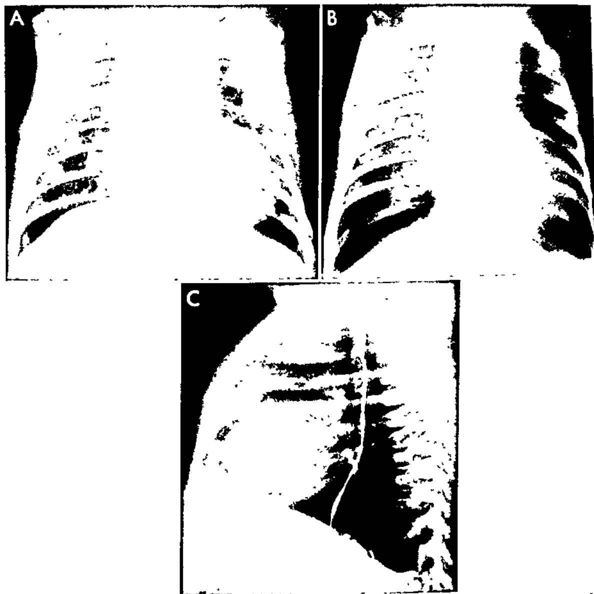


Fig. 226a.—Pulmonary atresia Girl, aged 2½ months (F H. 570608). The apex is slightly upturned and rounded Great enlargement of right atrium The main trunk and main branches of the pulmonary artery cannot be identified Great reduction in vascularity of lungs

mally in weight. The *cardiac findings* were characterized by an accentuated but pure second sound and a very faint systolic murmur The *electrocardiogram* showed signs of right atrial hypertrophy but not ventricular hypertrophy

Roentgenologic examination — Slight enlargement of the heart could be ascribed to dilatation of the right atrium The apex was upturned and, as in the preceding case, the left border of the ventricle was prominent (Fig 226a). The main trunk of the pulmonary artery was not visualized, the vascularity of the lungs was sparse, and the aorta was somewhat wide The picture was reminiscent of that in tetralogy of Fallot.

Cardiac catheterization —The catheter passed from the right atrium into a ventricle, lying anteriorly, and with a pressure of 132/10 mm Hg The oxygen saturation in this chamber was 30 per cent, that in the right atrium 35 per cent, and that in the superior and inferior vena cava 40 per cent Pressure recording in the right atrium showed giant *a* waves (13 mm Hg) and slightly raised mean pressure The left side of the heart could not be catheterized, and no arterial sample was obtained When the catheter tip lay in the right ventricle, a small quantity of contrast medium (test dose) was injected under fluoroscopic control A small cavity, about 2 by 2 cm, was filled, but the most striking feature was that

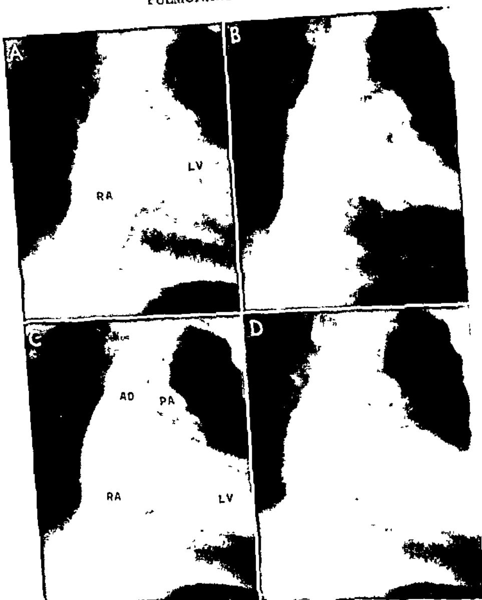


Fig. 226b —Same case as in Figure 226a The wall of the right ventricle is greatly hypertrophied Cavity (*RV*) very small The pulmonary artery (*PA*) is filled through a patent ductus arteriosus (arrow in F) The right atrium (*RA*) is very large; it communicates with the left atrium (*LA*) through a wide atrial septal defect The aorta (*AO*) is given off entirely from the left ventricle (LV) RAA, right auricular appendage (*continued*)

the contrast medium remained in the cavity, without being transported any farther Since the injection elicited ventricular tachycardia, the catheter tip had to be withdrawn into the right atrium, into which the final injection of contrast medium was made

Angiocardiography—Large parts of the heart were visualized simultaneously, which made it more difficult to interpret the anatomy, particularly that of the pulmonary orifice, which was not at first realized to be atresic. In retrospect, when the autopsy findings were available, the small right ventricle—which in

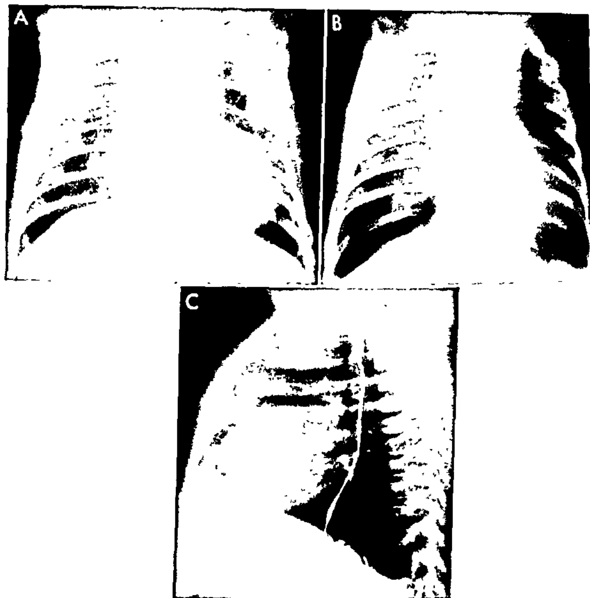


Fig. 226a.—Pulmonary atresia Girl, aged 2½ months (F.H 570608). The apex is slightly upturned and rounded Great enlargement of right atrium The main trunk and main branches of the pulmonary artery cannot be identified Great reduction in vascularity of lungs.

mally in weight The *cardiac findings* were characterized by an accentuated but pure second sound and a very faint systolic murmur The *electrocardiogram* showed signs of right atrial hypertrophy but not ventricular hypertrophy.

Roentgenologic examination.—Slight enlargement of the heart could be ascribed to dilatation of the right atrium The apex was upturned and, as in the preceding case, the left border of the ventricle was prominent (Fig 226a). The main trunk of the pulmonary artery

..

of Fallot

Cardiac catheterization—The catheter passed from the right atrium into a ventricle, lying anteriorly, and with a pressure of 132/10 mm Hg. The oxygen saturation in this chamber was 30 per cent, that in the right atrium 35 per cent, and that in the superior and inferior vena cava 40 per cent Pressure recording in the right atrium showed giant *a* waves (13 mm Hg) and slightly raised mean pressure. The left side of the heart could not be catheterized, and no arterial sample was obtained When the catheter tip lay in the right ventricle, a small quantity of contrast medium (test dose) was injected under fluoroscopic control A small cavity, about 2 by 2 cm, was filled, but the most striking feature was that

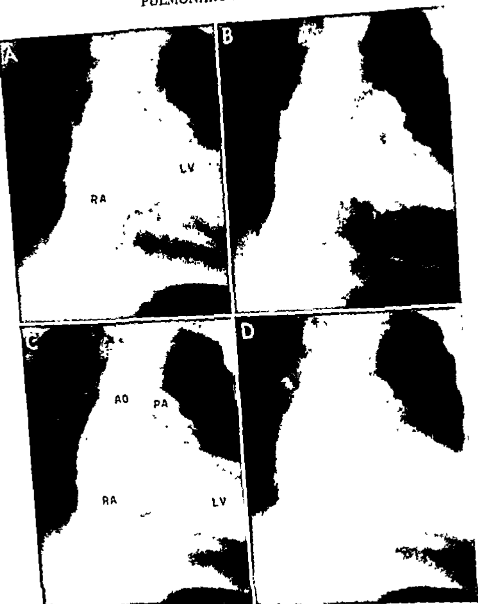


Fig 226b —Same case as in Figure 226a The wall of the right ventricle is greatly hypertrophied Cavity (*RV*) very small The pulmonary artery (*PA*) is filled through a patent ductus arteriosus (arrow in *F*) The right atrium (*RA*) is very large, it communicates with the left atrium (*LA*) through a wide atrial septal defect The aorta (*AO*) is given off entirely from the left ventricle (*LV*) *RAA*, right auricular appendage (*continued*)

the contrast medium remained in the cavity, without being transported any farther Since the injection elicited ventricular tachycardia, the catheter tip had to be withdrawn into the right atrium, into which the final injection of contrast ...

particularly that of the pulmonary orifice, which was not at first realized to be atresic In retrospect, when the autopsy findings were available, the small right ventricle—which in ...

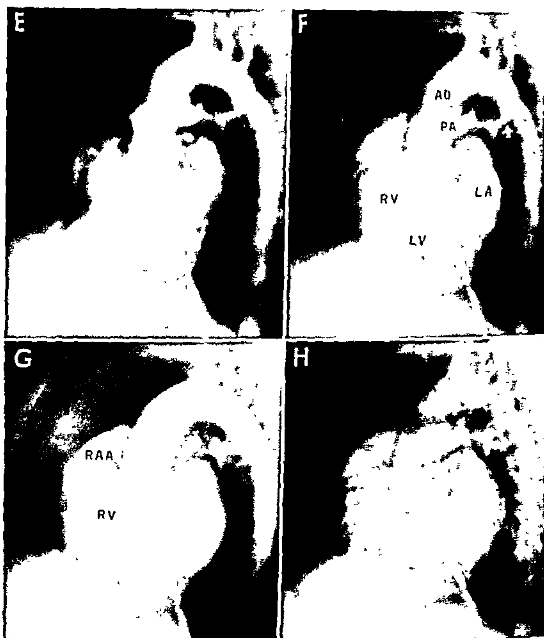


Fig. 226b (*cont.*)

tum was possible, owing to superimposition of the large right atrium

Autopsy.—Two weeks after examination, the patient died at the local hospital. At autopsy, the pulmonary orifice was found to be atresic and the main trunk of the pulmonary artery narrow. The other atrioventricular valves were normal. The ductus arteriosus was patent, and the foramen ovale was wide. The ventricular septum was intact and the wall of the right ventricle hypertrophied.

A feature of interest is that a distinct bulge in the left superior border of the heart on the roentgenograms could be ascribed in one case to dilatation of the infundibulum and superior part of the sinus region of the right ventricle (Figs. 225a and b). In the other case the bulge was due to hypertrophy of the left ventricle (Figs. 226a and b). The cardiac outline had the appearance usually stated to be characteristic of tricuspid atresia (p. 700). This illustrates the difficulty of using this particular feature as a basis for a differential diagnosis.

In both cases it should have been possible to make a correct diagnosis based on the findings at catheterization and angiocardiography, but deficient knowledge of this type of malformation led to misinterpretation of the findings. In our series, such high pressure in a venous chamber as in these two cases had not occurred at such an early age except in exceedingly severe pulmonary stenosis or atresia. Nor had retention of contrast medium in an enclosed cavity, which lacked a vascular appearance, been seen in other cases. In our opinion, injection of a small quantity of contrast medium into the venous chamber, under fluoroscopic control, is practically decisive for the diagnosis. This test injection should determine the subsequent angiocardiographic procedure. If the ventricle is small, the final injection of contrast medium should be made into the right atrium; if it is of ordinary size, the medium may be injected into the right ventricle.

In both these cases cardiac catheterization was incomplete, but even if all the chambers of the heart had been catheterized, the diagnosis could not have been established without angiocardiographic examination, for severe pulmonary stenosis with a large right to left interatrial shunt may present the same features at catherization.

8 Stenosis or Atresia of Pulmonary Artery Branches

STENOSIS or atresia of the pulmonary artery branches is not infrequently present in combination with intracardiac malformations (231, 236, 494, 501). Considerably more cases would certainly be diagnosed if, at cardiac catheterization, the pressure were always recorded continuously on passage of the catheter from the PCV position to the main trunk of the pulmonary artery (Fig. 227). However, with this procedure, only those stenoses are detected which involve the branches explored by the catheter. At angiocardiography, on the contrary, the anatomy of the whole vascular tree is depicted, so that all the branches can be evaluated. It can therefore be expected that increasing use of angiocardiography will lead to more frequent demonstration of these malformations.

Emanuel and Pattinson (236) stated, on the basis of reports in the literature and experience in their own cases, that in combination with malformation of the bulbus cordis, there is absence of the *left* pulmonary artery branch alone. On the other hand, if the heart is normal or malformations not involving the bulbus cordis are present, it is usually the *right* pulmonary artery branch that is absent. This statement does not agree with the findings in our series.

We have observed 14 cases of stenosis or atresia of one or both main branches of the pulmonary artery in combination with various intracardiac malformations. Thus atresia of the left branch was present in one case of tricuspid atresia, one of mitral atresia, one of transposition of the great vessels, and one of tetralogy of Fallot. Atresia of the right branch was observed in two cases of tetralogy of Fallot and one of truncus arteriosus communis. Stenosis of the left branch was present in one case of tetralogy of Fallot Stenosis of the right branch was observed in two cases of valvular pulmonary stenosis, two cases of valvular and infundibular stenosis, and one case of supravalvular pulmonary stenosis. Finally, stenosis of both branches was found in one case of valvular pulmonary stenosis. Angiocardiographic studies were made in every case. Poststenotic dilatation was invariably demonstrated.

Stenosis at the site of origin of the main branches may be caused by a constricting pericardium or be associated with an extension of fibrous bands from the ligamentum arteriosum (619)

As an isolated malformation, stenosis (311, 490) or atresia (7, 259) of the pulmonary artery branches is rare. Hypoplasia of the main trunk of the pulmonary artery alone is also uncommon (703). Our series contains one case of a membranous impediment in the main trunk slightly distal to the valve Since the clinical features were identical with those of valvular pulmonary stenosis, this case has been assigned to the relevant section (p. 222).

Multiple stenoses of the peripheral ar-

teries of the lung, in all probability of congenital nature, seem to have been described first by Arvidsson, Möller, and Karnell (21, 23, 490) A familial occurrence was present in two of their cases, as in two cases reported by Van Epps (665).

According to the literature, roentgenologic examination in such cases shows the pulmonary trunk and its main branches to be hypoplastic, of ordinary width or dilated The central vessels have been described as irregular in several cases, and occasionally as aneurysmally dilated. The peripheral vessels have been evaluated as of ordinary width or narrow Signs of hypertrophy or

case was classified as primary pulmonary hypertension. Dr. G. Jonsson suggested to us the presence of multiple peripheral stenoses; this could be confirmed by a further analysis of the angiocardiograms. The salient features in these cases were as follows.

BOY, AGED 6 YEARS (C.G. 490706).—He had had frequent respiratory tract infections Since he started to walk he had been easily fatigued on exertion; he was particularly troubled by dyspnea, but had never been cyanotic. His development was normal. The heart sounds were normal A grade 3 systolic murmur was audible over the second left intercostal space and was transmitted to the left axilla The *electrocardiogram* was normal.

Cardiac catheterization.—The catheter

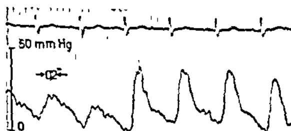


Fig 227.—Stenosis of the right pulmonary artery branch Girl, aged 17 (Case 78/56) with atrial septal defect Pressure recorded *on withdrawal* of the catheter from the right branch of the pulmonary artery to the main trunk.

possibly dilatation of the right ventricle have also been observed It has been stressed that in many cases the peripheral branches in the hilum are wider than the central ones, so that a tentative diagnosis can be made at *ordinary* roentgenologic examination (311). Experience particularly in cases studied by selective angiocardiography (23, 311) has shown that the degree and shape of stenosis vary greatly. All transitions have been observed between slight indentation of the vessel and atresia, and between *membranous* and elongated stenosis

Our series includes *two* cases In one, stenoses were observed *in* the branches of the left pulmonary artery Considerable stenosis of the right main branch prevented visualization of its branches, *which* could not therefore be evaluated. In the other case the peripheral stenoses were present bilaterally In the *first* edition of this book, this

passed through *the left* pulmonary artery

atrium 1 mm Hg The catheter passed through a patent foramen ovale *into the* left atrium (pressure 4 mm Hg), left *ventricle (81/6),* and pulmonary vein In *the PCA position* in the right lung, no typical arterial pulse curve was recorded, but a *damped* curve with a mean pressure of 7 mm Hg. *No shunts* were present The arterial oxygen saturation was 97 per cent

Roentgenologic examination.—The vascularity in the right *lung* was greatly reduced, and both the *central* and the peripheral branches were very *narrow.* In the left lung, the vessels were dilated and both the central and the perihilar branches were

228) It showed the presence of

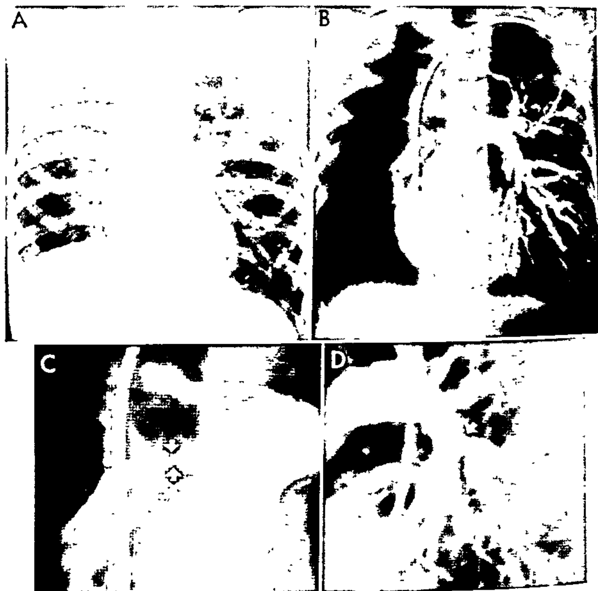


Fig. 228.—Stenosis of pulmonary artery branches Boy, aged 6 (C G 490706) Marked stenosis of right branch of pulmonary artery, about 2 cm distal to bifurcation (arrows point to stenosis) Distal to the stenosis, the pulmonary artery branches are very narrow. Normal branching on left side Some irregularity in outline of occasional branches and stenosis can be observed

multiple stenoses and poststenotic dilatation of several vessel branches in the left lung, considerable stenosis of the right pulmonary artery, and minimal central branches

Girl, aged 4 years (M J 490620).—The parents had not noticed any symptoms until the past year A murmur had been detected on a routine examination Since then, she had [illegible]

faint continuous murmur was audible round the whole thorax The *electrocardiogram* showed right ventricular hypertrophy

Cardiac catheterization —The pressure in the right ventricle was 82/4 mm Hg The catheter could not be advanced into the pulmonary artery (it was introduced through the saphe- [illegible]

passed through a patent foramen ovale, and a right to left interatrial shunt was demonstrated, with an oxygen saturation of 97 per cent in the pulmonary venous blood and 89 per cent in the arterial blood

Roentgenologic examination —The main trunk of the pulmonary artery was slightly di-

Angiocardiography —Contrast medium was injected into the right ventricle It showed considerable variations in caliber in the main trunk and main branches of the pulmonary artery in systole and diastole, as well as multiple stenoses and suggested poststenotic dilatation of the branches in the hilum and peripheral to it on both sides (Fig 229).



Fig 229.—Stenosis of pulmonary artery branches. Girl, aged 3 (M.J. 490620) Stenosis of practically all the pulmonary artery branches on both sides, and poststenotic dilatation of several branches (arrows point to most marked stenoses).

The presence of a continuous murmur in peripheral stenoses of the pulmonary artery branches has been described by Moller (490) and has also been demonstrated in experimentally induced stenosis (231) In our first case, there was a pure systolic murmur It was localized to the left side and was evidently not caused by the stenosis. Judging by the findings on angiocardiography, the flow through the stenosis must have been extremely small, and the murmur due to it therefore had so little energy that it was inaudible at this great depth.

Multiple stenoses of the pulmonary artery branches produce such a great increase in resistance that a rise in pressure occurs in the right ventricle, together with hypertrophy. Atresia or severe stenosis of only one main branch does not, on the contrary, result in any marked rise in pressure at rest. During exercise, however, there is reason to expect a greater rise than normal, owing to the reduced vascular bed. No exercise tolerance test could be made in our cases Because of the increased respiratory dead space, the ventilatory work can be anticipated to be greater than normal

9 Pulmonary Stenosis with Ventricular Septal Defect and Right to Left Shunt (Tetralogy of Fallot)

THE TETRALOGY of Fallot is a combination of: (1) pulmonary stenosis, (2) subaortic ventricular septal defect, (3) over-riding aorta, and (4) hypertrophy of the right ventricle due to the three malformations. Their origin is dependent on the evolution of the truncus-conus ridge (see p. 38). This association of anomalies gives rise to a characteristic picture, its degree of severity varying considerably with the anatomy of the malformations. The degree of pulmonary stenosis is the chief decisive factor. Thus, the more severe the stenosis, the greater the diminution in pulmonary blood flow and the larger the shunt. The difficulty of evaluating the degree of over-riding of the aorta has been discussed in connection with the description of the anatomy of ventricular septal defects (p. 339). The borderline between a tetralogy of Fallot and pulmonary stenosis with normal aortic root is indefinite. Severe pulmonary stenosis with a ventricular septal defect may be associated with a large right to left shunt and a clinical picture identical to that of tetralogy of Fallot, even when the aortic root is in normal position (Fig. 127, p. 140). In our series, we have therefore classified such cases under tetralogy of Fallot (see the classification of pulmonary stenosis, p. 138). Additional variations in the signs and symptoms are caused by the development of collateral branches to the pulmonary circulation.

The most extreme degree of pulmonary stenosis is atresia. According to Taussig (650), cases both of pulmonary stenosis and of atresia were included in the description given by Fallot in 1888. Tetralogy of Fallot is often used to denote cases of stenosis only, whereas those of atresia have been denoted as "pseudotruncus" or as "truncus aorticus" (650). Functionally, the difference is merely one of degree. It may sometimes be impossible on clinical examination to determine whether or not there is any blood flow through the pulmonary orifice. In such cases, the clinical picture is entirely dependent on the size of the pulmonary flow maintained by a patent ductus arteriosus or by collaterals from the bronchial arteries. Clinically, a true truncus arteriosus with aplasia of both branches of the pulmonary artery, the lungs being supplied only through the bronchial arteries, cannot be distinguished from tetralogy of Fallot with pulmonary atresia, closed ductus arteriosus and collateral circulation through the bronchial arteries. The distinction is embryologic and anatomic and is of no clinical importance. Consequently, we have chosen to classify both these types as tetralogy of Fallot with pulmonary atresia (see p. 330).

As a rule, the pulmonary stenosis is infundibular, the whole infundibulum being constricted into a long, narrow channel. It may, however, be confined to the ostium infundibuli, in which case a third ventricle of variable size is formed between the stenosis and the valve. Valvular stenosis alone

is an infrequent finding, whereas combined valvular and infundibular stenosis is common (see further p. 283).

The foramen ovale is remarkably often patent in tetralogy of Fallot, according to Abbott, this applied in 29 of 73 cases (cit. Edwards (216)). There is sometimes a wide interatrial communication which, under certain conditions, may also permit a large right to left shunt In about 25 per cent of all cases there is a right-sided aortic arch (216, 465)

From the hemodynamic viewpoint, the essential factor is a shunt from the right ventricle to the aorta, which results in arterial oxygen unsaturation, a decrease in pulmonary blood flow, and an increase in systemic flow As a compensation for the anoxia, polycythemia develops. Bing *et al* (58) and Graf and Mannheimer (465) found oxygen *consumption decreased* in ...ch patients, whereas Morse and Cassels ...92) found it to be normal. In the most ...vere cases, the patients die soon after ...rth, but when pulmonary stenosis is mild, ...ey may live until middle age The most ...ommon cause of death in the *former cate*...ory is cerebral anoxia, and in the milder ...ases, with survival for a longer time, death ... usually due to heart failure, bacterial ...ndocarditis, cerebral abscess, or cerebral ...hrombosis

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Our series includes 63 cases of tetralogy of Fallot. Dextrocardia was present in four of them One patient had atresia of the left main branch of the pulmonary artery and stenosis *at the site* of origin of the right branch. Pulmonary atresia was present in seven cases *Briefly*, these seven cases presented *the* following additional features.

BOY, AGED 8 YEARS (*L R.* 460604) —There was a patent ductus arteriosus, as well as a wide bronchial artery, which maintained ...

... per cent The diagnosis was established by cardiac *catheterization* and angiocardiography. The main trunk of the pulmonary artery was filled with contrast medium only from the ductus arteriosus and was atresic at the orifice.

BOY, AGED 2 MONTHS (R N. 530327).—The only source of blood supply to the lungs was through poorly developed bronchial arteries The diagnosis was based on cardiac catheterization and angiocardiography. Cyanosis was severe The case might also have been one of truncus arteriosus with aplasia of both branches of the pulmonary artery. Only autopsy could have permitted a differential diagnosis

GIRL, AGED 15 MONTHS (E B, 510822).—The pulmonary circulation received its blood supply only through the bronchial arteries Cyanosis was severe Angiocardiography was *not* performed. Autopsy disclosed atresia of the pulmonary artery.

BOY, AGED 4 MONTHS (B L. 520819).—The blood supply to the pulmonary artery took place through a patent ductus arteriosus. An atrial septal defect was also present. The diagnosis was made by cardiac catheterization and angiocardiography, and was verified at autopsy six months later. It was then found that, in addition, a pulmonary vein opened into the coronary sinus.

BOY, AGED 10 YEARS (M C. 470601) —The pulmonary circulation was supplied only *through collaterals from systemic arteries* On angiocardiography, *neither the main trunk* of the pulmonary artery nor its main *branches* were filled with contrast medium The aorta took its origin ...

... the patient was not cyanotic.

BOY, AGED 6 YEARS (A V. 510904) —On angiocardiography, both main branches of the pulmonary *artery* were filled from systemic collaterals, but *the* ...
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CLINICAL FEATURES

Retarded physical development is characteristic of all congenital heart diseases with pronounced cyanosis (465, 650). Fig-

9 Pulmonary Stenosis with Ventricular Septal Defect and Right to Left Shunt (Tetralogy of Fallot)

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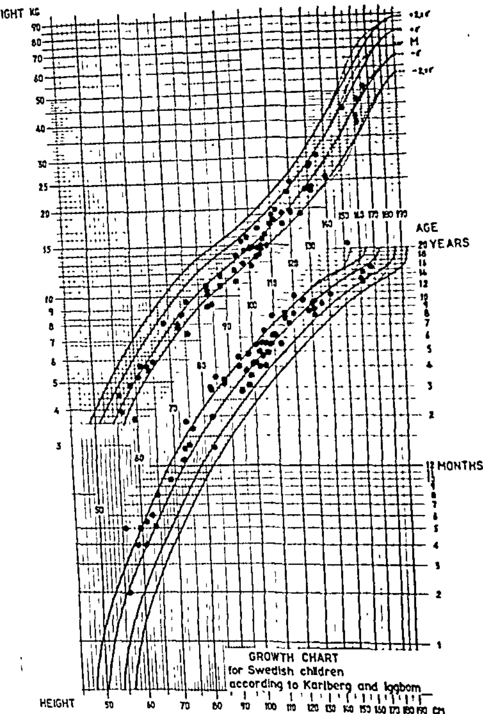


Fig 230 —Physical development of 63 children with tetralogy of Fallot Stunting of growth is evident (The relevant growth chart was kindly placed at our disposal by Drs P. Karlberg and S Iggbom [374, 375])

ure 230 shows the age distribution and physical development in our cases. Stunting of growth is evident.

Cyanosis is the most prominent symptom. In the most severe cases, it is noticeable from birth, but it may sometimes appear only after several years, when physical exertion is greater. The degree of cyanosis is dependent on the absolute quantity of reduced hemoglobin in the capillary blood and not only on arterial oxygen saturation (445), consequently, it becomes more marked when polycythemia develops. There is sometimes no increase in hemoglobin concentration. In such cases, hemoglobin saturation of the blood corpuscles is low, and there is reason to suspect iron deficiency (492, 542). If iron is administered,

TABLE 5.—ONSET OF CYANOSIS IN 62 CASES OF TETRALOGY OF FALLOT*

	Age			
	0–1 Mo	1–11 Mo	1–3 Yr	4–7 Yr
No of Cases	23	23	13	3

*One patient aged 5 months, with pseudotruncus was not cyanotic

polycythemia develops. The hemoglobin metabolism is raised (542). One of us (363) has found that the red blood corpuscles have a shortened survival time (60 to 80 days).

Many of our patients did not exhibit cyanosis until after they were 1 year old (Table 5). Only 1 was acyanotic at the time of examination. Cyanosis is usually the reason why congenital heart disease is suspected. In 19 of our patients the disease was detected in the course of a routine medical examination, when a murmur was heard. Hourglass nails and clubbing of the fingers and toes were present in all but seven children over 1 year of age, and a high hemoglobin concentration (between 15 and 26 Gm per 100 cc) in all but nine, four of whom were less than 5 months old Dyspnea on exertion was present in 38 cases

Spells of unconsciousness and sometimes convulsions are more common during the first year of life, they had occurred in 19 of our patients. A squatting position was adopted by 35 of the children; 13 of them were so young that this symptom could not be evaluated. Taussig (650) pointed out the importance of this symptom in patients with an inadequate pulmonary blood flow and stated that "all these children are able to get their breath more easily in this position." According to Lurie (446), the venous return to the heart is improved in the squatting position, and this explains the relief it affords. Brotmacher (104, 105) found an increase in systemic flow confined to the upper part of the body with an increase in oxygen tension. The arterial oxygen saturation returned more rapidly to the resting level after exercise if the patient was in the squatting position.

PHYSICAL WORKING CAPACITY.—As a result of the right to left shunt, the oxygen transport of the circulatory system is ineffective, and the physical working capacity is therefore low The effective stroke volume, i e., the quantity of blood per heart beat which passes from the pulmonary to the systemic circulation, is small The arterial oxygen saturation falls during exertion, and the low oxygen tension in the tissues limits the patient's ability to carry out physical work. Cerebral anoxia may even become so severe that unconsciousness supervenes. This does not necessarily imply that the oxygen-transporting ability of the circulation has reached its maximum. We have observed cases in which the arterial oxygen saturation fell during exercise to 30 to 40 per cent and the patient was unable to continue, despite a pulse rate no higher than 130 per minute In some cases, on the other hand, the exercise tolerance test could be continued until the pulse rate had reached about 170 per minute, the arterial oxygen saturation then having fallen to 40 to 50 per cent

Since most of our patients were small children, it was not as a rule possible to make an exercise tolerance test. The degree of disablement was therefore evaluated only on the basis of the history and of the observations during hospitalization. Thirty-three patients were severely disabled (un-

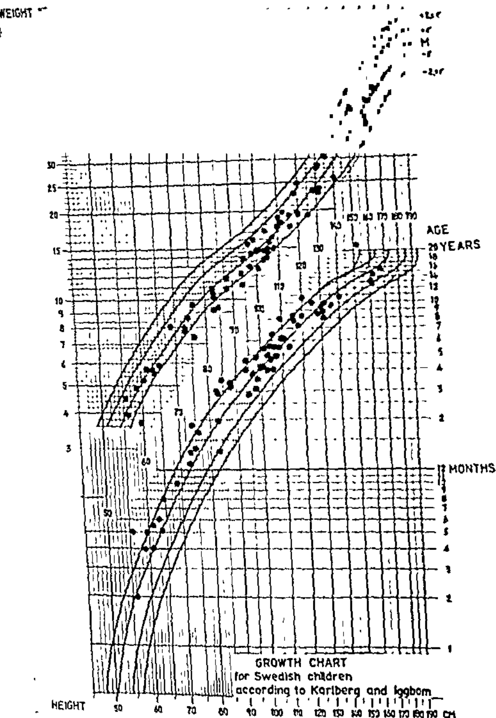


Fig 230 — Physical development of 63 children with tetralogy of Fallot Stunting of growth is evident (The relevant growth chart was kindly placed at our disposal by Drs P Karlberg and S Iggbom [374, 375].)

able to walk more than 100 meters on the level). This group includes 16 infants who were referred to hospital at this early age owing to the severity of their symptoms. Moderate disability was present in 17 cases (able to walk 100 to 1,000 meters) and slight disability in 14 cases (able to walk for some distance but lacking normal freedom of movement). Some improvement in the condition is often found after the first year of life, in that the severe attacks cease. On the other hand, both the degree of disablement and the cyanosis become more apparent when the patient has started to walk and thereby undergoes greater strain.

In two cases a Blalock-Taussig operation had been performed earlier. Although the anastomosis functioned, the flow was inadequate and both patients were still deeply cyanotic.

Physical Signs

Palpation of the precordium.—Larsson and Mannheimer (465) and Métianu and Durand (202) have stressed that precordial heave is common, but the former authors have also emphasized that this phenomenon is presumably due in many cases to a malformation of the thorax of noncardiac origin. In our series, this symptom was marked in five cases, and a slight precordial heave was noted in 11 cases. This contrasts to a certain extent with the roentgenologic findings of a normal-sized heart in most cases of tetralogy of Fallot, but can be explained on the grounds of the marked right ventricular hypertrophy. As a result, the apex beat was faint or not palpable in all our cases, whereas a distinct parasternal lift was observed in 15 cases.

Sounds and murmurs.—The *first heart sound* is normal and is most distinct over the apex (554). Varying descriptions have been given of the *second heart sound*. Some authors (107, 554) have stated that it is weak or absent over the pulmonary area. Others (36, 202, 650, 713), have pointed out that it is often heard most distinctly and may even be accentuated over the second left interspace. The second sound is generally regarded to be caused by the closing of the aortic valve. This can easily be demonstrated on the phonocardiogram. When the aorta is over-riding, the valve lies closer to the anterior thoracic wall than normally, and the flow through the aorta is increased. The flow through the pulmonary artery, on the contrary, is reduced, the pressure is low and the orifice often lies more dorsally than normal (see Angiocardiography, p. 295). The sound phenomenon most characteristic of tetralogy of Fallot is therefore the pure second sound, which usually is accentuated over the pulmonary area (Fig. 231). In nine of our 54 cases in which the phonocardiogram was recorded, it was nevertheless possible to observe a faint pulmonary component with a splitting varying between 0.02 and 0.08 sec (Fig. 232, *A*). It was, however, so faint that it was not audible on auscultation. In their series, Leatham and Weitzman (418) found that the pulmonary component did not appear until after a Blalock-Taussig operation or after valvulotomy.

The *systolic murmur* is not uniform, with regard to the intensity, to the position in the pulse period or to the site on the precordium at which it is loudest. This is not surprising, in view of the facts brought forward with respect to the systolic murmur in various types of pulmonary stenosis with normal aortic root (see p. 146). In tetralogy of Fallot, there is often combined valvular and infundibular stenosis. The narrow channel into which the infundibulum is converted is presumably of greater importance for the systolic murmur (202, 650). Since there is a ventricular septal defect, there is no difficulty in emptying the right ventricle. Consequently, systole is not prolonged, and the cardiac output through the infundibular or valvular stenosis becomes pathologically decreased. This explains Brown's observation (107) that the systolic murmur is often weaker in tetralogy of Fallot than in "isolated" pulmonary stenosis.

A systolic murmur was present in 57 of our 63 cases. In 42, it was heard and re-

corded most distinctly in the third or fourth left interspace and in 15 over the pulmonary area. In 18 cases the murmur was classified as loud, in 33 as moderate, and in six as faint. No systolic murmur could be detected in six cases. In three cases, a continuous murmur was audible on both sides of the thorax, owing to large bronchial ar-

corded. Figure 232, C, is a phonocardiogram in a case of valvular pulmonary stenosis in tetralogy of Fallot, the patient was a 5-year-old boy. The murmur was late systolic (see p. 147). Figure 232, D, shows a continuous murmur recorded over the second right interspace. The patient, a 7-year-old boy, had been operated on six months earlier and

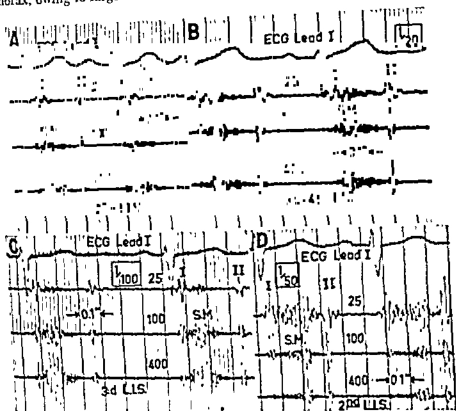


Fig. 231.—Phonocardiograms in four cases of tetralogy of Fallot. *A*, boy, aged 3 (B.S. 500122). *B*, boy, aged 5 (L.L. 470607). *C*, boy, aged 6 (G.R. 460930). *D*, boy, aged 5 (K.J. 480422). In all, infundibular stenosis was shown on angiocardiograms. Note early systolic murmur. The aortic component of the 2nd sound has large amplitudes, no pulmonary component. Boxed figures denote degree of amplification, other figures denote standard frequencies of the filters. *L.I.S.*, left interspace, *SM*, systolic murmur.

teries. In typical cases, the murmur was early systolic, i.e., that characteristic of infundibular stenosis, this is evident from Figure 231. Exceptions were, however, found. Figure 232, D, [illegible]

loud murmur during the whole of systole, but, surprisingly enough, no definite continuous murmur could be either heard or re-

showed great improvement. The continuous murmur was a definite indication of a functioning anastomosis.

Thus, in tetralogy of Fallot, a moderately loud, often early systolic murmur is heard, usually most distinctly in the left third interspace. A strong or accentuated second sound, which is always pure and not split, is audible over the base, to the left of the sternum. Neither a precordial bulge nor a

able to walk more than 100 meters on the level). This group includes 16 infants who were referred to hospital at this early age owing to the severity of their symptoms. Moderate disability was present in 17 cases (able to walk 100 to 1,000 meters) and slight disability in 14 cases (able to walk for some distance but lacking normal freedom of movement). Some improvement in the condition is often found after the first year of life, in that the severe attacks cease. On the other hand, both the degree of disablement and the cyanosis become more apparent when the patient has started to walk and thereby undergoes greater strain.

In two cases a Blalock-Taussig operation had been performed earlier. Although the anastomosis functioned, the flow was inadequate and both patients were still deeply cyanotic.

Physical Signs

Palpation of the precordium.—Larsson and Mannheimer (465) and Météanu and Durand (202) have stressed that precordial heave is common, but the former authors have also emphasized that this phenomenon is presumably due in many cases to a malformation of the thorax of noncardiac origin In our series, this symptom was marked in five cases, and a slight precordial heave was noted in 11 cases. This contrasts to a certain extent with the roentgenologic findings of a normal-sized heart in most cases of tetralogy of Fallot, but can be explained on the grounds of the marked right ventricular hypertrophy As a result, the apex beat was faint or not palpable in all our cases, whereas a distinct parasternal lift was observed in 15 cases.

Sounds and murmurs —The *first heart sound* is normal and is most distinct over the apex (554). Varying descriptions have been given of the *second heart sound* Some authors (107, 554) have stated that it is weak or absent over the pulmonary area. Others (36, 202, 650, 713), have pointed out that it is often heard most distinctly and may even be accentuated over the second left interspace The second sound is generally regarded to be caused by the closing of the aortic valve. This can easily be demonstrated on the phonocardiogram. When the aorta is over-riding, the valve lies closer to the anterior thoracic wall than normally, and the flow through the aorta is increased. The flow through the pulmonary artery, on the contrary, is reduced, the pressure is low and the orifice often lies more dorsally than normal (see Angiocardiography, p. 295). The sound phenomenon most characteristic of tetralogy of Fallot is therefore the pure second sound, which usually is accentuated over the pulmonary area (Fig. 231). In nine of our 54 cases in which the phonocardiogram was recorded, it was nevertheless possible to observe a faint pulmonary component with a splitting varying between 0.02 and 0.08 sec (Fig. 232, *A*). It was, however, so faint that it was not audible on auscultation. In their series, Leatham and Weitzman (418) found that the pulmonary component did not appear until after a Blalock-Taussig operation or after valvulotomy.

The *systolic murmur* is not uniform, with regard to the intensity, to the position in the pulse period or to the site on the precordium at which it is loudest. This is not surprising, in view of the facts brought forward with respect to the systolic murmur in various types of pulmonary stenosis with normal aortic root (see p. 146). In tetralogy of Fallot, there is often combined valvular and infundibular stenosis The narrow channel into which the infundibulum is converted is presumably of greater importance for the systolic murmur (202, 650). Since there is a ventricular septal defect, there is no difficulty in emptying the right ventricle. Consequently, systole is not prolonged, and the cardiac output through the infundibular or valvular stenosis becomes pathologically decreased. This explains Brown's observation (107) that the systolic murmur is often weaker in tetralogy of Fallot than in "isolated" pulmonary stenosis.

A systolic murmur was present in 57 of our 63 cases In 42, it was heard and re-

parasternal lift is uncommon. Variations in the findings on palpation and auscultation may be noted; they are most frequently due to complicating anomalies or to a less characteristic manifestation of the disease. A diagnosis of tetralogy of Fallot based on physical examination alone is presumably only a probable diagnosis.

ELECTROCARDIOGRAPHY

The electrocardiogram in tetralogy of Fallot is characterized mainly by hypertrophy of the right ventricle. Although the changes are not as marked as those that may be seen in pulmonary stenosis with intact ventricular septum, they are found more consistently. The QRS complex typical of right ventricular hypertrophy, with a tall R wave and a small S wave to the right over the precordium and a small R wave and a deep S wave to the left, is almost invariably present in tetralogy of Fallot. The ECG was normal in only four of our cases The patients were between 2 and 5 months old, and right ventricular hypertrophy was not yet sufficiently developed to differ from the normal ECG in such young infants. An abnormally tall R and V_1 was present in all but the four infants just mentioned and in the cases with dextrocardia A tall R wave in VR was, on the other hand, less common. A prolonged ventricular activation time in V_1 was recorded in scarcely half of the cases and then amounted to 0 04 sec. Only in four cases was it as long as 0.06 sec

A right bundle-branch block was found in none of our patients, but a first degree A-V block with a P-Q interval measuring 0.26 sec was present in one case In contrast to pulmonary stenosis with intact ventricular septum, the T waves in this group of patients were not pathologic Abnormally high P waves in V_1 were found about as often as in pulmonary stenosis with intact ventricular septum

As a rule, the heart was slightly rotated clockwise [illegible]

[illegible] was present in four cases

In seven patients with a right to left interatrial shunt, the ECG changes were the same as in the uncomplicated cases. One patient (L.R. 460604) with pulmonary atresia had a highly developed collateral circulation through both the bronchial arteries and the ductus arteriosus; this resulted in such an increase in pulmonary blood flow that arterial oxygen saturation was 83 per cent. In this case, the electrocardiogram gave evidence not only of right ventricular hypertrophy but also of left ventricular strain.

ROENTGENOLOGIC EXAMINATION

In the majority of cases of tetralogy of Fallot, the roentgenologic appearance is typical The variations are nevertheless great and are essentially to be ascribed to the diversity of the anatomic conditions in the outflow tract of the right ventricle and the pulmonary artery and to the associated modifications in hemodynamics. Thus, the conditions are complicated, and it is by no means in every case that the roentgenologic examination provides sufficient information to assess the degree of severity. Although clinically mild cases usually exhibit only relatively slight changes (Fig 233), the lack of conspicuous roentgenologic abnormalities does not rule out the presence of considerable disability (Fig. 234).

The most common roentgenologic features in our 71 cases of tetralogy of Fallot were the following

An enlarged right ventricle, often with a bulging anterior surface and upturned apex.

A concavity in the cardiac border at the site of the infundibulum

[illegible] the lungs

Enlargement of the right atrium and dilatation of the aorta were also found in many cases.

With a coeur en sabot, the aforementioned changes were extremely marked. This type was observed in the younger age groups in particular.

The typical outline of the heart in the frontal view is to be attributed chiefly to the

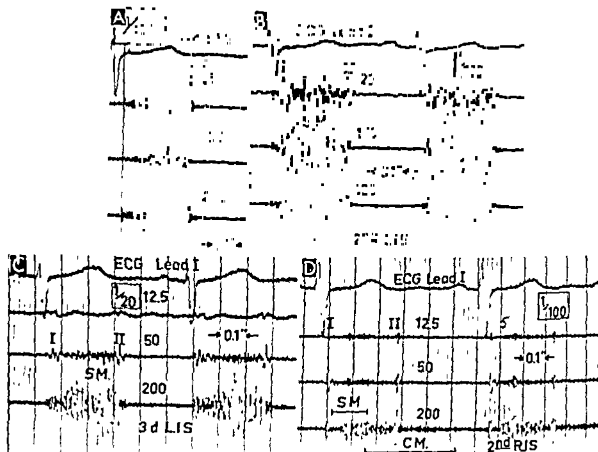


Fig. 232.—Phonocardiograms in cases of tetralogy of Fallot Boxed figures denote degree of amplification, other figures denote standard frequencies of filters *C M.*, continuous murmur, *L.I.S* and *R I S*, left and right interspaces, *S M*, systolic murmur *A*, with infundibular and mild valvular pulmonary stenosis Girl, aged 1 (K.K 550918). Note pulmonary component of second sound 0 07 sec after aortic component *B*, with large patent ductus arteriosus Boy, aged 8 (Y.G. 440424). Note loud systolic murmur filling whole of systole *C*, with slight infundibular and marked valvular pulmonary stenosis. Boy, aged 5 (L H 470204). Note late systolic murmur *D*, with continuous murmur due to functioning anastomosis after Blalock-Taussig operation Boy, aged 7 (G R 460930).

abnormal conditions in the infundibular region, combined with rotation of the heart and enlargement of the sinus region of the right ventricle, which produces tip-tilting of the apex (621, 688). The backward and upward displacement of the apex (Fig. 235) is accentuated by the smallness of the left ventricle (226, 583). The hypertrophy and enlargement of the right ventricle are most easily evaluated in the lateral view; they are then apparent as an increased anterior bulging of the often enlarged anterior surface of the heart toward the thoracic wall (Fig 235). This was observed in 45 of our cases The curvature of the border of the right ventricle is considerably more marked in tetralogy of Fallot than in pulmonary stenosis with normal aortic root In the oblique projection, one of the factors which makes it difficult to judge the size and shape of the right ventricle [illegible] in the lateral view.

A shallow indentation in the cardiac border at the site of the infundibulum is often distinct (Fig 236). it was visible in 42 of *our cases.* A comparison with the angiocardiographic findings shows that this part of the cardiac border corresponds to the site *of the stenosis* (Fig. 297). This is also the reason for the retraction of the heart in the anterosuperior part of the right ventricle, as seen in the lateral projection (Figs. 235, 236 and 242). This effect may be partly masked by the overlapping of the right auricular appendage. In the frontal or oblique projection, slight bulging of the cardiac border above the indentation is not infrequently seen, it is caused by the infundibular chamber (Figs 236 and 237). The left auricular appendage sometimes forms part of the border in this region, seen [illegible] lar region can always be visualized free from the overlapping left auricular appendage and descending aorta On fluoroscopy an inward movement of the infundibulum

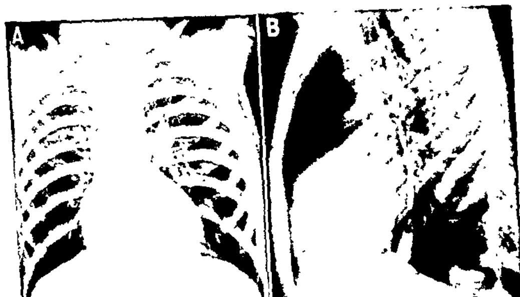


Fig 234.—Tetralogy of Fallot Boy, aged 5 (T.A 481028) Severely disabled Slight but somewhat more marked roentgenologic findings than in Figure 233 Hypertrophic shape of right ventricle with curvature of anterior surface, inappreciable upturning of apex, main trunk of the pulmonary artery not visible, moderate reduction in vascularity; slight dilatation of aorta, enlargement of right atrium

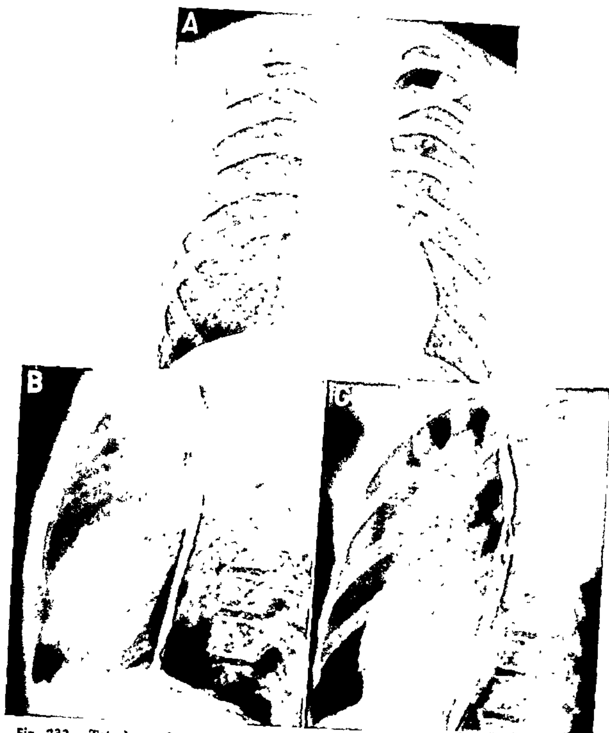


Fig. 233.—Tetralogy of Fallot Boy, aged 15 (B Ö 371008) Very slightly disabled Only slight hypertrophy of right ventricle, *no upturning* of apex but distinct dilatation of aorta, ordinary prominence of pulmonary artery, normal vascularity of lungs

abnormal conditions in the infundibular region, combined with rotation of the heart and enlargement of the sinus region of the right ventricle, which produces tip-tilting of the apex (621, 688). The backward and upward displacement of the apex (Fig. 235) is accentuated by the smallness of the left ventricle (226, 583). The hypertrophy and enlargement of the *right ventricle* are most easily evaluated in the lateral view; they are then apparent as an increased anterior bulging of the often enlarged anterior surface of the heart toward the thoracic wall (Fig 235) This was observed in 45 of our cases The curvature of the border of the right ventricle is considerably more marked in tetralogy of Fallot than in pulmonary stenosis with normal aortic root In the oblique projection, one of the factors which makes it difficult to judge the size and shape of the right ventricle is the ventral displacement of *the ascending* aorta, due to overriding This factor has no disturbing effect in the lateral view.

A shallow indentation in the cardiac border at the site of the infundibulum is often distinct (Fig. 236); it was visible in 42 of our cases A comparison with the angio- [illegible] reason for the retraction of the heart in the anterosuperior part of the right ventricle, as seen in the lateral projection (Figs. 235, 236 and 242). This effect may be partly

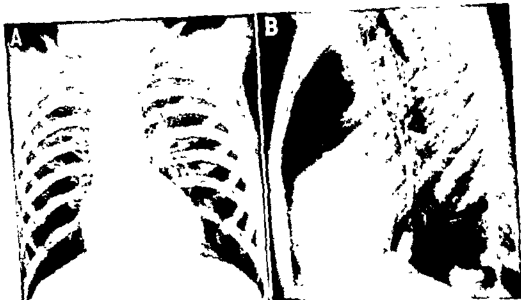


Fig 234.—Tetralogy of Fallot. Boy, aged 5 (T A 481028). Severely disabled. Slight but somewhat more marked roentgenologic findings than in Figure 233 Hypertrophic shape of right ventricle with curvature of anterior surface, inappreciable upturning of apex, main trunk of the pulmonary artery not visible, moderate reduction in vascularity, slight dilatation of aorta, enlargement of right atrium

masked by the overlapping of the right auricular appendage. In the frontal or oblique projection, slight bulging of the cardiac border above the indentation is not infrequently seen, it is caused by the infundibular chamber (Figs. 236 and 237). The left auricular appendage sometimes forms part of the border in this region, seen in the frontal view, as may be inferred from the autopsy findings (Fig 235). In the right anterior oblique projection, the infundibular region can always be visualized free from the overlapping left auricular appendage and descending aorta. On fluoroscopy, an inward movement of the infundibulum



Fig. 235.—Tetralogy of Fallot Girl, aged 1 year (G-B A. 520620), see Figure 273 (p 293) Great hypertrophy of sinus region of right ventricle, spherical anterior surface and marked up-turning of apex Arrow in *E* points to a shallow indentation, corresponding to the borderline between the right and the left ventricle, as seen in the autopsy specimen (*F*) Borderline between

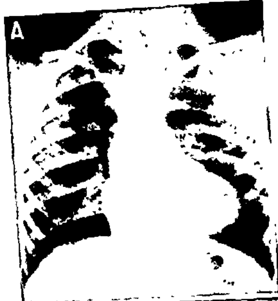




Fig. 236.—Tetralogy of Fallot Boy, aged 4 (L P. 491023), see Figure 297 (p 318). Considerable hypertrophy of sinus region of right ventricle and marked upturning of the apex. C, dilated, right sided aorta displaces trachea to the left and makes a slight indentation in it immediately above the bifurcation, greatly decreased prominence of pulmonary artery, local prominence at the site of the pulmonary artery is caused mainly by the third ventricle (upper arrow) (see Fig 297), lower arrow marks site of infundibular stenosis Great reduction in vascularity, vascular markings in lungs are partly reticular, probably representing collateral circulation

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the ventricles is identified by the anterior descending branch of the left coronary artery Left ventricle is displaced upward and rests like a cap on the large sinus region The roentgenogram also shows that the left auricular appendage almost entirely overlaps the left segment of the pulmonary artery Aorta wide and aortic arch right-sided, narrow pulmonary artery, considerable reduction in vascularity of lungs, enlargement of right atrium AO, aorta, LAA, left auricular appendage, *LV* and RV, left and right ventricles, PA, pulmonary artery, *RA*, right atrium, *SVC*, superior vena cava.



Fig. 235.—Tetralogy of Fallot Girl, aged 1 year (G-B A. 520620), see Figure 273 (p 293). Great hypertrophy of sinus region of right ventricle, spherical anterior surface and marked upturning of apex. Arrow in *E* points to a shallow indentation, corresponding to the borderline between the right and the left ventricle, as seen in the autopsy specimen (*F*) Borderline between

. . ... In

kind is illustrated in Figures 238 and ... (p 290); the wall of the third ventricle was thin, and inverse pulsations with systolic expansion were visible.

The lack of normal prominence of the pulmonary artery (Figs. 235 and 236) is due partly to its small caliber and partly to its abnormal course, obliquely and medially into the mediastinum. A comparison of roentgenologic, angiocardiographic, and markedly decreased, unless an extensive collateral circulation entirely or partly compensates for the decreased blood volume in the pulmonary circulation. It is best evaluated by correlating the fluoroscopic observations—when the main object is a study of the size of the pulsations—with the findings in roentgenograms made at short exposure times Even finer branches of the pulmonary vessels appear distinctly in such pictures (Figs 235, 236, 239, 240). Pulsations in the hilar vessels can seldom be seen on fluoroscopy. Movements in the hilar regions



Fig. 239 —Tetralogy of Fallot with typical coeur en sabot appearance. Boy, aged 2 (V S 510415) Marked curvature of anterior outline of right ventricle, pulmonary artery not visible greatly reduced vascularity

electrokymographic examinations shows that, as a rule, only a small, basal segment of the artery is outlined against the lung.

Consequently, because of the unfavorable conditions applying to the projections, it is seldom possible to draw direct conclusions from the roentgenologic examination with respect to the size of the pulmonary artery Angiocardiography shows that poststenotic dilatation of the pulmonary artery may be present, although it is not visible on the roentgenogram. Only exceptionally can a distinct "pulmonary arc" be identified, representing slight but definite dilatation of the main trunk.

The vascularity of the lungs usually is

may be caused by impacts from the dilated aorta.

Dilatation of the aorta was observed in one-third of our cases (Figs. 234, 236, 241-243) The dilatation is caused by the increased blood flow It is not directly related to the degree of ...

position in the sense of a displacement of the aortic orifice to the right (202, 659, 722) In tetralogy of Fallot, the ventricular ...

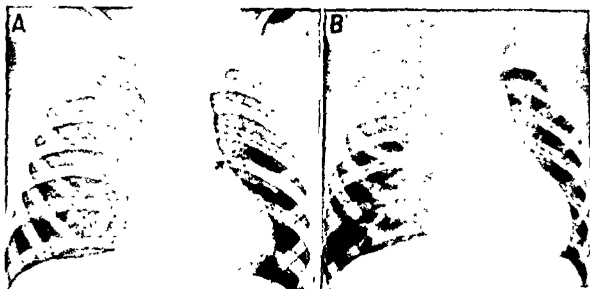


Fig. 237.—Tetralogy of Fallot Boy, aged 4 (S.J. 480315). At arrow, a small local prominence corresponding, according to findings on angiocardiography, to a third ventricle. Vascularity of the lungs, which was considerably reduced before operation according to Blalock *(A)*, has increased postoperatively *(B)* No change in outline of heart.



Fig. 238.—Tetralogy of Fallot with infundibular and valvular stenosis Boy, aged 9 (Y G ... trunk of the ... is dis-

quently, the actual implication of dextroposition is, in fact, a forward displacement of the aortic root over the right ventricle. Selective angiocardiography shows that the aortic orifice is consistently displaced more or less ventrally to its normal site, but not to the right. In several of our cases, the displacement was, in fact, slightly to the left instead, it was presumably caused by the typical rotation of the heart about a vertical axis

A right to left interatrial shunt was present in four cases The roentgenologic findings differed in no respect from the typical picture in tetralogy of Fallot. Thus, there was no dilatation of the left atrium, and no enlargement of the left ventricle could be demonstrated. In three of the cases, upturning of the apex was conspicuous.

In the slightly enlarged right atrium, the appendage is often prominent, an increase in presystolic activity may be seen on fluoroscopy and can be recorded on the electrokymogram

It is often difficult to estimate the size of the left ventricle by roentgenologic examination. When a notch is present at the apex, it constitutes the borderline between the right and the left ventricle (Figs. 235 and 240). Eck (226) has pointed out that, in the left oblique projection, the small left ventricle may sometimes appear as a cap that lies behind and above the right ventricle.

In the majority of our cases, there was no pathologic increase in the heart volume; when enlargement was found, it was only moderate. In the total heart volume, a diminution in volume of the left atrium and left ventricle compensates for the enlargement of the right ventricle and right atrium. Moreover, the enlargement of the right ventricle is never gross.

In some cases of tetralogy of Fallot, no definite branches of the pulmonary artery in the hilar region or in the periphery of the lungs, on one or both sides, can be identified either at fluoroscopy or on the roentgenograms. This applies in pulmonary atresia in particular. The vessels in the lungs emanate wholly or partly from a collateral circulation which is distinctly reticular, it is usually most extensive in the vicinity of



Fig 241.—Tetralogy of Fallot with infundibular and valvular stenosis Boy, aged 7 (L-K. 460207); see Figure 278 (p. 299). Heart has typical *coeur en sabot* shape, main trunk of the pulmonary artery cannot be identified, its central branches are narrow, vascularity of the lungs is reduced. Vascular markings, particularly basally, have distinctly reticular appearance, representing extensive collateral circulation. Aorta is wide.

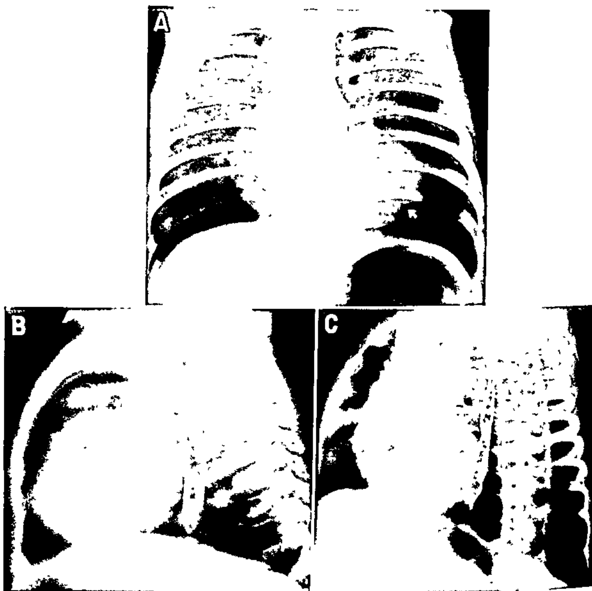
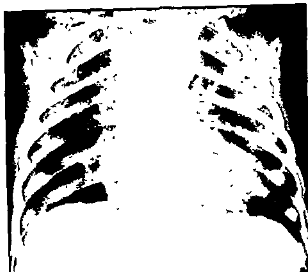
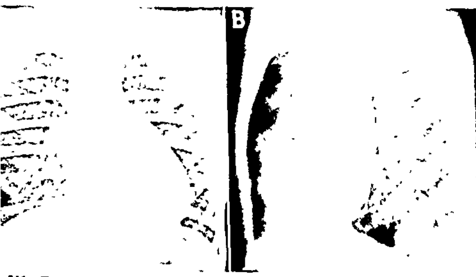


Fig. 240.—Tetralogy of Fallot Girl, aged 4 months (S S. 520419); see Figure 293 (p. 314). Heart has typical coeur en sabot shape, greatly reduced vascularity. pulmonary artery not visible, large right atrium, marked notch at apex (arrow in *A*), indicating border between ventricles At age 6 weeks, changes were barely discernible, except for definite decrease in vascularity.



[2]43.—Tetralogy of Fallot Boy, aged 3 (B S. 500122), see Figure 288 (p 309). Aorta wide, on the left extending as far as the clavicle, main trunk of pulmonary artery not great reduction in vascularity



244 —Tetralogy of Fallot Boy, aged [illegible]

the hilum and basally (Figs. 241, 242 and 244). The individual elements in this reticular arrangement have a stippled appearance. In some of our cases, a number of tortuous vessels were seen in the basal part basal part of the lungs was present in several of these cases, it may have been due to the passage of the collaterals through the pleura (Fig. 244, *D*). Changes in vascular markings characteristic of a collateral cir-



450903);
ngs,
s of

of the left lung, they possibly originated in the phrenic artery (Fig. 245). The collateral circulation may sometimes be so highly developed that one may be inclined to conclude that the pulmonary circulation is increased (Fig 244).

Thickening of the pleura around the culation were observed in 17 cases Mediastinal vessels, belonging to the collateral circulation, which cause a bulge in the esophagus have been described (156, 226), but were encountered in only one of our cases.

In pulmonary atresia, some of the varia-



Fig 245.—Tetralogy of Fallot. Girl, aged 5 (M.V 481009), see Figure 290 (p 311). Contrast-filled collaterals are visible, they originate in the dilated left inferior phrenic artery (between arrows)

A

B

C

Fig 246 —Pseudotruncus Girl, aged 15 months (E B. 510822). Extremely scanty vascularity both centrally and peripherally, with reticular appearance throughout, owing to the large thymus, aorta is difficult to evaluate At autopsy, aorta was found to originate almost entirely in right ventricle, bronchial arteries were wide

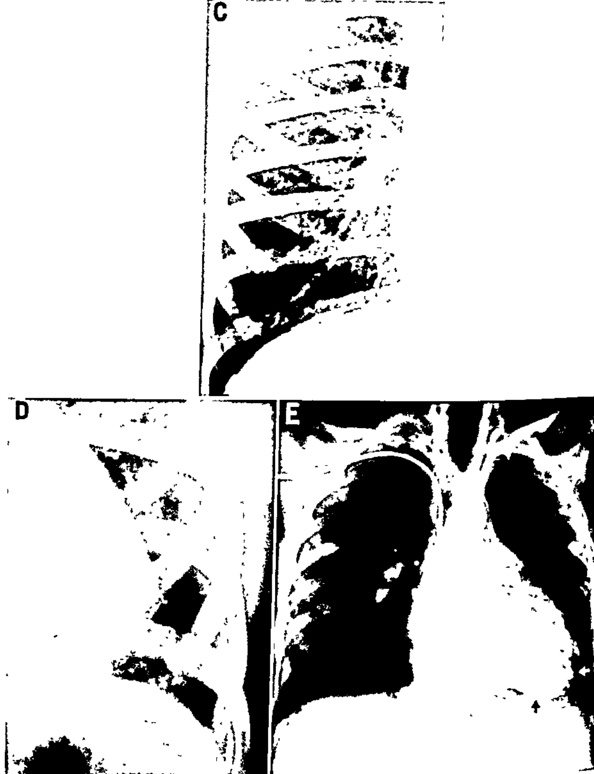


Fig. 244 (*cont.*).—*C*, vascularity of lungs is increased and has distinctly reticular appearance *D*, pleural thickening, possibly also due to the extensive collateral circulation, part of which is filled at angiocardiography (arrows in *E*) A fine net of collaterals is seen in the left flank, and thicker branches at the left base


A


B
C



Fig. 247.—Pseudotruncus. Boy, aged 2 months (R N. 530327); see Figure 301 (p 324) Right ventricle is grossly hypertrophic, apex is upturned; vascular pedicle narrow, vascularity of lungs considerably reduced

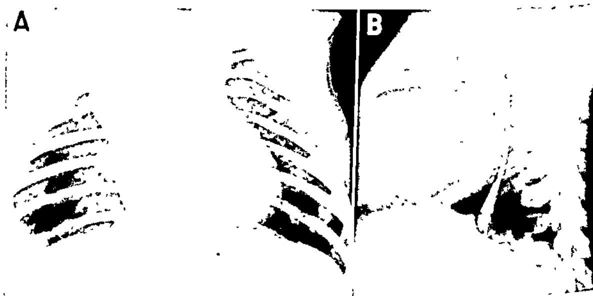
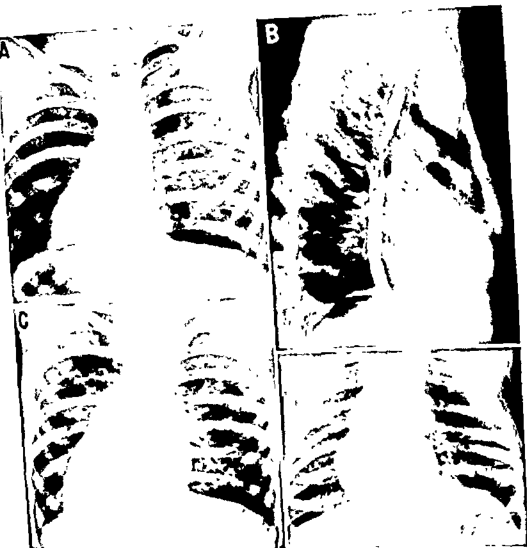


Fig. 248.—Pseudotruncus Boy, aged 4 months (B L 520819), see Figure 299 (p. 320) Apex slightly upturned, conspicuous expansion of superior part of mediastinum to right caused by dilated, right-sided aorta, greatly reduced vascularity of lungs, vascular markings have reticular appearance.



Figs 251 and 252 —Tetralogy of Fallot with infundibular and valvular stenosis

Fig 251 (*above* and *below left*) —With mirror-image dextrocardia Girl, aged 5 (M P 481020), see Figure 274 (p 295) Bulge in superior part of right outline is caused by a small third ventricle C, definite increase in vascularity postoperatively

Fig. 252 (*below right*) —With rotation of the heart Girl, aged 11 (A M H. 421218) Right outline of heart is formed by the right ventricle, which overlaps right atrium and lies anterior to it Aorta is wide and bulges considerably to right Marked reduction in vascularity. Vascular markings have a reticular appearance, especially basally

tions found in the roentgenogram are associated with the development of a collateral circulation. Consequently, they are conditioned to some extent by age. During infancy, the roentgenologic appearance is often very typical; it is characterized by extremely sparse vascularity of the lungs and by considerable enlargement and hypertrophy of the right ventricle. Marked concavity of the outline in the infundibular region is often seen, but is not as constant as the other features. If there is a plentiful blood supply through collaterals or a patent ductus, the vessels are dilated, particularly in the hilum, and their pulsations can sometimes be observed. These variations are illustrated in five of our seven such cases (Figs. 246-249), their clinical features are described on page 249. Cardiac catheterization was performed in *all* seven and angiocardiographic examination in six of them An autopsy was made on two patients In six cases the vascular markings were notably sparse, indicating scanty pulmonary blood flow In all but one case the right ventricle was enlarged and the apex upturned. The aorta was greatly dilated in three of these cases, whereas it was somewhat on the narrow side in the others None of them presented a *conspicuous coeur en sabot.* In the remaining case (Fig 249 the pulmonary circulation was supplie through the ductus arteriosus and bron chial arteries. In this case, pulsations we seen in the hilum, and there was no dim nution of pulmonary vascularity. The le atrium was slightly dilated, and angi cardiography showed the left ventricle t be as large as the right. The aorta was di tinctly dilated. The outline of the apex wa normal.

The incidence of a right-sided aortic arc is slightly over ·20 per cent in large seri (202, 226). This anomaly was present eight of our cases. In two of them the ar ran unusually far in the cephalic directio and in two the left subclavian artery ori nated at an abnormal site and caused typical impression in the esophagus fro behind *(Fig. 250) Other* vascular ano alies, such as a persisting communicati between the left superior vena cava and t right atrium, were also observed. Dext cardia or anomalies of rotation other th the characteristic, moderate backward tation of the apex were observed in fo cases (Figs 251-253).

Complicating pulmonary lesions are n uncommon. In two of our cases the lun *were emphysematous and cystic.* The de

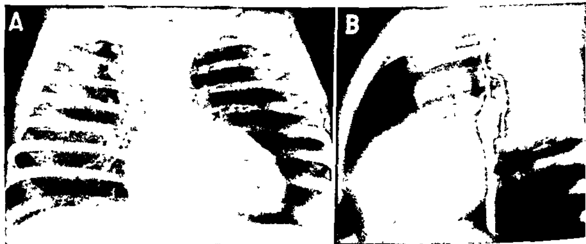


Fig. 250.—Tetralogy of Fallot Boy, aged 8 months (B B. 520329), see Figures 277a and 27 (pp 298 and 299) *Aorta long and wide and arch right-sided,* descending aorta runs downwa on right side Left subclavian artery passes behind esophagus, causing indentation in it Sm bulge in superior part of left outline (arrow) is formed by narrow pulmonary artery, which greatly displaced to left Vascularity is considerably reduced.

onstration of such pulmonary lesions is of great importance as far as the indications for operation are concerned, since the prospect of obtaining satisfactory results is then considerably poorer. Decreased lung volume with an underdeveloped hemithorax were observed in the absence of one pulmonary artery (Figs. 254a and 254b).

ELECTROKYMOGRAPHY

The abnormal course of the pulmonary artery often makes it difficult, and sometimes impossible, to obtain technically satisfactory electrokymograms of this vessel

Eighteen cases were studied, tracings from the pulmonary artery were obtained in 16 of them Both these and the electrokymograms of the infundibular region were of the same type as in infundibular stenosis with ventricular septal defect but without a right to left shunt (Figs 255 and 256). With predominating valvular stenosis, the pulmonary artery tracings exhibited features typical of both valvular and infundibular stenosis

In one case in which the third ventricle was unusually large (see p 261, and the angiocardiogram, Fig. 271), inverse pulsations were recorded over the greater part of this ventricle (Fig 257). They were of the same type as those which in other cases occurred only in the vicinity of the valvular plane.

In some cases, the tracing from the infundibular region suggested that a short segment of the outline was formed by the left auricular appendage.

In the aortic electrokymogram, the incisura and dicrotic wave were, as a rule, ill defined and were situated far down on the descending limb (Fig. 258). This may have been dependent on the large blood flow in the dilated aorta, as well as on the drainage through the collaterals Such alterations were particularly conspicuous in the recordings of a case of pulmonary atresia and a wide patent ductus arteriosus (Fig. 259), the tracings from the pulmonary artery in this case were indicative of a highly abnormal circulation

An electrokymogram of the right atrium indicating an impediment to its emptying was recorded in a few of the cases (Fig 260)



Fig 254a —Tetralogy of Fallot Boy, aged 11 (H O. 440229) Right main branch of pulmonary artery fairly wide The left main branch is lacking Left side of thorax smaller than right







[illegible] of pulmonary orific[illegible]
line [illegible]
s wi[illegible]
med[illegible]
cann[illegible]
be identified in any projection There is no definite reduction in vascularity, but it has an abnormal, reticular, stippled appearance. Site of the stomach also anomalous

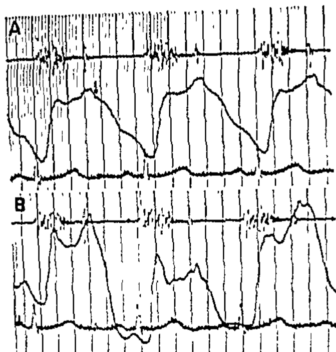


Fig 255 —Electrokymograms of pulmonary artery and infundibular region in tetralogy of llot (valvular and infundibular stenosis). Boy, aged 8 (E R. 460930) PCG over apex A, pulonary artery: early upstroke, horizontal course in systole, reduced incisura (arrow) and diotic wave B, infundibulum, close to valve systolic expansion due to superimposed artery



Fig. 254b —Same case as in Figure 254a Moderate constriction of ostium infundibuli (*OI*) and of infundibulum of right ventricle (right-hand *I*) Infundibulum of left ventricle (left-hand *I*) somewhat wide Circular constriction (between arrows in *D*) of right main branch of pulmonary artery, slightly after its origin from the main trunk Left main branch lacking *Left* side of thorax underdeveloped *PA*, pulmonary artery, *PB*, parietal band, *RV*, right ventricle

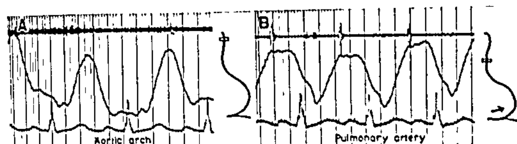


Fig 259 —Electrokymograms of aortic arch and pulmonary artery in pulmonary atresia (pseudotruncus) and wide patent ductus arteriosus Boy, aged 8 (L.R. 460604). PCG· in *A*, over aortic area (continuous murmur), in *B*, over apex Tracing of aortic arch has same appearance as in aortic incompetence. In pulmonary artery tracing, rise is slow and continuous until end of systole, and incisura and dicrotic wave are situated high up

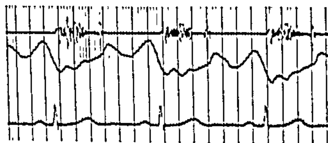


Fig 260 —Electrokymogram of right atrium in tetralogy of Fallot, same case as in Figure 255. PCG over apex Deep presystolic deflection of long duration (0 10 sec) Emptying phase in early diastole is shallow

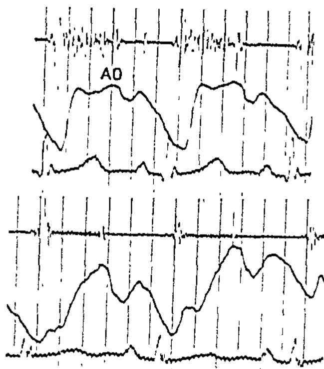


Fig. 256a (*above*).—Electrokymogram of pulmonary artery in tetralogy of Fallot (infundibular and valvular stenosis). Girl, aged 10 (B.S 450902). PCG over apex. *AO*, aortic component of 2nd sound. Plateau in systole. Descending limb starts at aortic component and ends with typical segment. Distinct dicrotic wave

Fig 256b (*below*).—Electrokymogram of pulmonary artery in tetralogy of Fallot with predominating valvular stenosis Boy, aged 10 (S.H. 451227) PCG over apex. Basal anacrotic notch and slow secondary systolic rise Descending limb with terminating typical segment Distinct dicrotic wave.

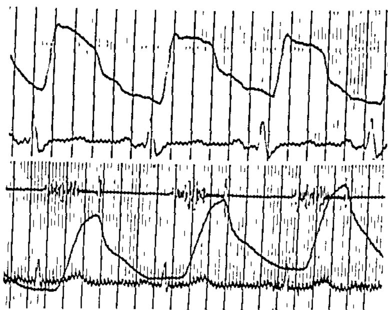


Fig. 257 (*above*).—Electrokymogram of third ventricle in tetralogy of Fallot Boy, aged 8 (Y G 440424) Systolic dilatation of third ventricle

Fig. 258 (*below*)—Electrokymogram of aortic arch in tetralogy of Fallot, same case as in Figure 255. PCG over apex Incisura and dicrotic wave are almost obliterated and are far down on descending limb

lacks the typical appearance of an arterial pulse curve and shows small, irregular deflections and respiratory variations (Fig 261). It is sometimes possible to judge the anatomic conditions of the stenosis on the basis of the withdrawal curve. This matter has been dealt with in more detail on page 182

In 16 cases the catheter passed from the right ventricle into the aorta, and in five, into the left ventricle. In 15 cases it passed from the right to the left atrium, and in one case to the left ventricle by this route In an additional case in which the left atrium was not entered, a large atrial septal defect was found at autopsy; the patient died in the course of operation It is there-

the right atrium is high, the pressure is usually higher in the former. In our series, systolic pressure (the *a* wave) in the right atrium averaged 6 mm Hg; in only five cases was it as high as 10 mm Hg. Thus, the rise in pressure in the right atrium is not as marked as in severe pulmonary stenosis with intact ventricular septum. In tetralogy of Fallot, a right to left interatrial shunt can occur only under certain conditions Of our 15 cases in which the catheter passed from the right to the left atrium, a right to left interatrial shunt was present in seven.

All these seven patients had severe pulmonary stenosis or atresia, and the pressure in the left atrium was extremely low

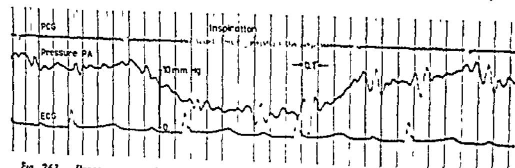


Fig 261 —Pressure in pulmonary artery in tetralogy of Fallot with infundibular stenosis Girl, aged 5 (I C 490401) Curve lacks typical appearance of an arterial pulse curve, exhibiting small, irregular deflections and marked respiratory variations in pressure

fore probable that a patent foramen ovale or an atrial septal defect is present in a considerably larger number of cases than is disclosed b...

eve

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tec ...que, it is, however, more difficult to pass through the foramen ovale than when a leg vein is used

A right ...

of

of

pa ...ood (713) found no such shunt in three cases in which the left atrium was catheterized, and Lagerlof *et al* (465) made the same observation in four cases Although the blood flow through the left atrium is abnormally low and that through

Severe pulmonary stenosis with a scanty collateral circulation results in a small reflow to the left atrium and a pressure lower than in the right atrium In one of the cases, a pulmonary vein opened into the coronary sinus, this caused a further decrease in reflow to the left atri...

crease

favore...

teratria. shunt Obviously, a shunt of this kind is found with a complicating stenosis of the tricuspid valve, and such a case is described on page 731. The eight cases with no interatrial shunt presented the features of uncomplicated tetralogy of Fallot. Thus, a right to left interatrial shunt is associated with tetralogy of Fallot when there is a complicating tricuspid malformation, or when there is very severe stenosis with an

CARDIAC CATHETERIZATION

The first reports of cardiac catheterization in cases of tetralogy of Fallot (60, 242) were able to elucidate the hemodynamic mechanism in this disease. Since then, the method has been used as a routine diagnostic test (292, 465), even though it is regarded by many workers as superfluous. It is true that in typical cases the diagnosis can be established on the basis of the clinical picture, including the electrocardiogram and roentgenograms, but in some cases the evaluation is difficult. The diagnostic value of cardiac catheterization in such cases as these is, however, limited by the following factors.

A diagnosis of tetralogy of Fallot can be made with the help of cardiac catheterization only if the catheter passes from the right ventricle into both the aorta and the pulmonary artery. The systolic pressure in the right ventricle is the same as that in the aorta, whereas it is low in the pulmonary artery. Pulmonary stenosis is of such a degree that the shunt goes from the right ventricle and into the aorta Attempts to advance the catheter into the aorta or into the pulmonary artery are often unsuccessful This applies particularly to young infants, when the catheter must be inserted into a saphenous vein. Owing to the abnormal direction of the infundibulum of the right ventricle (see Fig 269, p 286), considerable difficulty may then be encountered in maneuvering the catheter into the pulmonary artery. In the presence of severe stenosis or atresia, this is obviously impossible

The question of a differential diagnosis applies first and foremost to pulmonary stenosis with intact ventricular septum and a right to left shunt through an atrial septal defect. The fact that the pressure in the right ventricle is on the same level as that of the systemic artery is of no decisive diagnostic value. This is because a pressure of this order of magnitude is strikingly often found in pulmonary stenosis with intact ventricular septum as well. If the catheter can be passed into the left atrium, the right to left shunt can be demonstrated Under certain conditions, such a shunt is also found in tetralogy of Fallot. Blood sam ples taken simultaneously from the left atrium and the systemic artery then show that a right to left shunt is present both be tween the atria and between the right ven tricle and the aorta.

Determination of the cardiac output ac [illegible]

especially to all cases with a right to left shunt. The blood flow through the pulmo nary artery is small, but since there is ofter an extensive collateral circulation througl the bronchial arteries, Fick's method can not be used to calculate the pulmonary flow a fact pointed out by Bing *et al.* (60) More over, in the youngest children, oxyge saturation of the blood in the individua chamber of the heart is apt to vary consider ably from one minute to the next All th blood samples cannot be taken simultane ously. Consequently, the best conception c the degree of severity of the disease is ob tained by observing the physical capacit of the patients, preferably by means o standardized exercise tolerance tests wit concurrent oximetric determinations

Cardiac catheterization was performe in all our cases. In our opinion, the exam nation should be made before the questic of operation is raised. The main reason that angiocardiography, with injection the contrast medium through the cathet directly into the right ventricle, can then made concurrently. This combined exam nation always confirms the diagnosis ar also gives a clear picture of the anatomy the right ventricular outflow tract, the ve tricular septal defect, the position of th aortic root, and the size and position of th great vessels and their branches, which m be a decisive factor in the choice of surgic technique (see p 288).

In 32 of our cases, the catheter pass into the pulmonary artery. The pressu was subnormal in every instance. An add tional decrease in the flow may be produc by the catheter. The curve not infrequent



Fig. 262.—*Tetralogy of Fallot* Considerable displacement of whole dextrodorsal conus ridge forward and to left, causing great constriction of infundibulum of right ventricle
the ridge (right part of crista supr
direction, bulging considerably int
Numerous pits and *channels* are
greatly thickened endocardium Se
muscular septum, lies to right of
dorsal to medial cusp of the tricus
the *defect* Great hypertrophy of th
fundibulum, *OI*, ostium infundib
septal defect
ventricular

extremely small reflow to the left atrium, or when one of the pulmonary veins opens at an abnormal site.

A shunt of this nature may also develop in right ventricular failure, but this is unusual in children. Esclavissat *et al.* (242) found a right to left interatrial shunt in some cases of tetralogy of Fallot, but they did not discuss its cause. Gasul *et al.* (271) described tetralogy of Fallot in a 9-year-old girl who died of heart failure. Roentgenologic examination disclosed considerable enlargement of the heart, and the ECG showed deviation of the electric axis to the left. Cardiac catheterization was not performed. Autopsy revealed tetralogy of Fallot and a patent foramen ovale 1.5 cm in diameter. The right atrium was enormously enlarged, but the tricuspid valve was not stenosed. In this case, there was evidently a right to left shunt due to cardiac failure. Microscopic examination showed myocardial necrosis, which may explain the heart failure.

HEMODYNAMICS DURING EXERCISE

In the presence of a large ventricular septal defect with or without over-riding aorta, the systolic pressure must be the same in the left and the right ventricle. Owing to the pulmonary stenosis, the pulmonary flow is restricted, but there is no impediment to emptying of the right ventricle into the aorta In pulmonary stenosis with intact ventricular septum, the stroke volume at rest can be maintained—up to a certain limit—during exercise as well, despite shortening of the ejection time This is possible on the grounds of the further rise in pressure in the right ventricle (see p 190).

In the pulmonary stenosis with a large ventricular septal defect or over-riding aorta, the pressure in the right ventricle cannot exceed that in the systemic arteries. A falling stroke volume in the pulmonary circulation could therefore be expected during exercise. Table 6 nevertheless shows that in tetralogy of Fallot (with 50 per cent over-riding aorta) it may remain fairly un-

TABLE 6.—HEMODYNAMICS AT REST AND DURING EXERCISE IN A PATIENT WITH TETRALOGY OF FALLOT*

Case No	Sex	Age, Year	Body Surface Area, m^2	Physical Working Capacity % of Predicted	Work Load, kg/min	O_2 Uptake, ml/min	Pulse Rate, beats/min	Pulmonary Circulation: AV O_2 Diff, ml/L	Pulmonary Circulation: Cardiac Output, L/min	Pulmonary Circulation: Stroke Vol, ml	Systemic Circulation: AV O_2 Diff, ml/L	Systemic Circulation: Cardiac Output, L/min	Systemic Circulation: Stroke Vol, ml	O_2 Cap., vol %	Art O_2 Sat, %	Shunt L/min: R → L†	Shunt L/min: L → R	Effective Stroke Vol, ml	Pressure, mm Hg: RA Mean	Pressure, mm Hg: RV Syst.	Pressure, mm Hg: PA Syst	Pressure, mm Hg: PA Diast.	Pressure, mm Hg: PA Mean	Pressure, mm Hg: Br. A. Syst.	Pressure, mm Hg: Br. A. Diast.	Pressure, mm Hg: Br. A. Mean
61/56	M	32	1.76	40	Rest	225	76	81	2.8	37	43	5.2	68	25.0	78	2.7	0.3	33	6	100	12	9	10	110	65	82
					200	746	128	167	4.5	35	69	10.8	84	25.0	56	6.4	0.1	34	9				9	127	64	87

*For abbreviations see Table 1, p 119 †Pulmonary venous blood assumed to be 97 per cent saturated

conus ridge run forward and often to the left, the parietal band being inserted cephalad (Fig 262). This produces narrowing of the infundibulum.

In normal cases, the infundibulum is *limited anteriorly* by the ventricular wall and posteriorly to the left by the ventricular

development of the dextrodorsal ridge, there is a diminution in the part of the anterior ventricular wall included in the conus

usually in the ostium infundibuli. In this case, the infundibulum often forms a separate chamber, corresponding to a third ventricle (Fig. 264). If, on the contrary, the parietal band is fused with the anterior and septal wall immediately below the valvular plane, severe stenosis develops (Fig 265) or possibly atresia (Fig 266). Distal to the stenosis or the atresic region, not infrequently normal (Fig 266) or *slightly thickened* valves (Fig. 267) are found at the usual site.

Small pits and channels are often present



Fig 264.—Tetralogy of Fallot *A*, anterior wall of the ventricle is dissected out to show right part of the course of the crista supraventricularis and its parietal band and insertion of the band, cranially and far to the left, slightly to right of the septum. Right part of the infundibulum, which lies above, is dissected out and raised to demonstrate position of the band and how it

septal defect.

formation and a proportionate increase in the part formed by the septum (Figs 263-265) The variations in development of these parts of the infundibulum are the cause of the variable appearance of the outflow tract When all the components of the conus ridge *deviate toward the septum*, the infundibulum is converted into an elongated, constricted *channel, corresponding* to tubular stenosis. When the anomaly is confined mainly to the right part of the crista supraventricularis and *its* parietal band, a circumscribed stenosis develops,

in the infundibulum and ostium infundibuli (Fig 262), they resemble those found in infundibular stenosis with normal aortic root. Endocardial thickening of the infundibulum narrows the lumen still more.

Infundibular stenosis is not uncommonly combined with valvular stenosis (Fig. 278). Various figures have been given for the incidence of valvular stenosis in tetralogy of Fallot Baffes *et al.* (30) gave a detailed account of the anatomic variations in the infundibulum and also of the incidence of valvular stenosis in an autopsy series of 42

changed, despite a considerable rise in the pulse rate. For, during exercise, the systolic pressure in the aorta rose, as in healthy individuals (344). The increase in the pulmonary flow during exercise was, however, smaller than the increase in the systemic flow; consequently, the right to left shunt must have increased. The arterial oxygen saturation fell from 78 per cent at rest to 56 per cent during exercise. The effective stroke volume was small under both conditions, as an expression of deficient oxygen transport. Quantitatively as well, this corresponded to the degree of decreased physical working capacity.

ANGIOCARDIOGRAPHY

MORPHOLOGIC BACKGROUND —In tetralogy of Fallot, there is a developmental anomaly in the infundibular region of the right ventricle. The site of the dextrodorsal conus ridge is abnormal When growth proceeds normally, this ridge contributes to formation of the septum and to closure of the interventricular foramen. Its lower part continues as the right boundary of the crista supraventricularis and its parietal band, which normally runs to the right in a caudal direction (Fig. 90, p. 86). In tetralogy of Fallot, these components of the

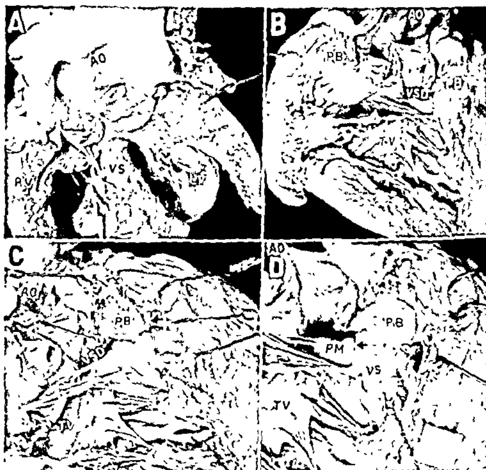


Fig. 263.—Tetralogy of Fallot Marked displacement of the dextrodorsal conus ridge, with considerable constriction of infundibulum, particularly of ostium infundibuli (arrow in C), the part above it is somewhat wider The way the wide aorta over-rides the ventricular septum is seen in A ... r septum Cusps hang down into ... es through the defect into the le ... ventricles. AO, aorta, I, infundit ... PM, papillary muscle, TV, tricus ... t



Fig 267.—Tetralogy of Fallot (pseudotruncus). Below the pulmonary orifice, dextrodorsal ... is fused with the ventricular septum, causing atresia of infundibulum Pulmonary



Fig 268.—Tetralogy of Fallot (pseudotruncus). Great displacement of dextrodorsal conus ridge ar
muscul
above
riding
points
right ventricles VS, ventricular septum



Fig. 265.—Tetralogy of Fallot. Dextrodorsal conus ridge is severed and anterior wall of the ventricle lifted to show cephalic insertion (*PB* on left). Third ventricle is small, and distance between stenosis and valvular plane short. Greatest constriction is at the level of the parietal band (ostium infundibuli). Above the band, the wall of the infundibulum is thin. *AO*, aorta, *OI*, ostium infundibuli, *PA*, pulmonary artery, *PB*, parietal band; *RV*, right ventricle; *3V*, third ventricle, *VS*, ventricular septum, *VSD*, ventricular septal defect.

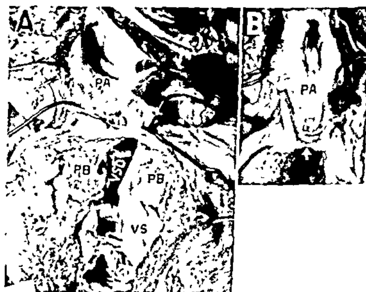


Fig. 266.—Tetralogy of Fallot (pseudotruncus) Dextrodorsal conus ridge is displaced so far cephalad and to the left that it is fused with the ventricular septum, thus causing atresia of the infundibulum Pulmonary artery is narrow Pulmonary cusps are normal, the atresia is caudal to them. Pulmonary artery is cut open to show the cusps (arrow in *B*) *PA*, pulmonary artery, *PB*, parietal band, *VS*, ventricular septum, *VSD*, ventricular septal defect



Fig 267.—Tetralogy of Fallot (pseudotruncus). Below the pulmonary orifice, dextrodorsal

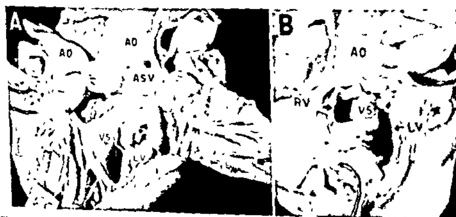
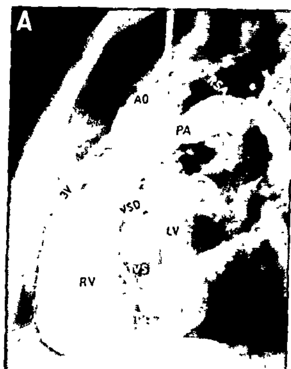


Fig 268.—Tetralogy of Fallot (pseudotruncus). Great displacement of dextro... ridge and atresia of infundibulum As usua... muscular part of septum which, in all our s... above VS in A, VS in B). Aortic cusps hang... riding being about 50 per cent (B, seen f... points to right ventricle AO, aorta (truncu... right ventricles, VS, ventricular septum.







F[illegible] conus ridge is greatly [illegible]ularly ostium infun- [illegible]d in Figure 264 The sinus region is large and forms the apex Superior, curved margin of the muscular part of the septum forms the lower limitation of the septal defec [illegible] has an irregular shape, with small pits and channels, which [illegible] Pul-monary artery is narrow and aorta wide, its degree of [illegible] ere is a slight leakage of contrast medium into the right atrium *A* and *F*, functioning anastomosis (Blalock's operation) between right subclavian artery and right branch of pulmonary artery.

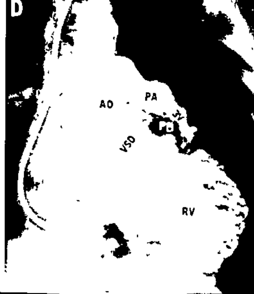




Fig 269 (cont.)

...ta, LV and RV, left and right ...
...conus ridge), R...
...entricular septal

children with tetralogy of Fallot; the majority were less than 3 years of age. Infundibular stenosis or atresia was present in every case. In nine cases there was a combination of infundibular and valvular stenosis; in no case was there valvular stenosis only. Brinton and Campbell (93) found, in a series with a different age distribution, that valvular stenosis alone was present in about one-fifth of the cases.

In tetralogy of Fallot, the ventricular septal defect is almost invariably large. It usually involves the whole membranous part of the septum and extends from the aortic valve down to the muscular part of the septum, of which the thick, curved margin forms the caudal limitation of the defect (Figs. 263, 264, and 268). The aortic root over-rides the ventricular septum to a varying degree.

Angiocardiographic examination.—In view of the surgical requirements, the angiocardiographic examination in tetralogy of Fallot should provide information about the following:

The anatomic conditions in the outflow tract of the right ventricle, with special consideration of the appearance of the infundibular stenosis and its distance from the valvular plane, as well as an estimation of the size of the third ventricle.

The diameter, course, and branching of the pulmonary artery and the presence of any valvular stenosis.

The over-riding of the aorta and an approximate determination of its degree

The anatomy of the systemic arteries and their position in relation to the pulmonary artery and its main branches

A prerequisite for a detailed analysis of the anatomy is that angiocardiography be performed in such a way that the regions of the heart and vessels in question are visualized, to the greatest possible extent, without overlapping and with optimal density. This implies that a selective examination must be made, by injecting the contrast medium directly into the right ventricle If intravenous injection is used, the aforementioned conditions cannot be entirely fulfilled This is because in one of the paired projections the contrast-filled right atrium overlaps certain parts of the outflow tract of the ventricle and makes the interpretation more difficult.

In our choice of projection, we have considered the frontal and lateral to be preferable to oblique projections for several reasons. The aorta and the systemic arteries, as well as the pulmonary artery and its branches, are best depicted in the frontal view. The ventricular septum is usually least distorted in the lateral view, and the over-riding of the aorta can then be established on purely anatomic grounds and the septal defect can be illustrated. The complicated anatomy of the infundibular region is easiest to survey in the frontal and lateral projections.

One drawback is that the superior part of the infundibulum and the bulb of the pulmonary artery overlap the aorta in the lateral projection, but this seldom causes any essential difficulties in the interpretation When the course of the main trunk of the pulmonary artery runs in an extremely medial direction, its posterior segment may be partly masked by the contrast-filled ascending aorta in the frontal projection The corresponding segment of the pulmonary artery is, however, seen without overlapping in the lateral view. If the catheter slips out of the right ventricle during the injection and the right atrium also becomes filled, the conditions are less easy to survey and the septal defect can seldom be identified.

We performed an angiocardiographic examination in 69 of our cases In all but one case, the stenosis or atresia of the infundibulum could be visualized; the patient was the first in our series to undergo this examination and the technique was unsatisfactory.

The appearance of the infundibular stenosis varies greatly. In most cases, the whole dextrodorsal conus ridge contributes to the constriction (Figs 269 and 270), and only exceptionally is the stenosis due essentially to the parietal band (Fig. 271). When the stenosis is circumscribed, the third ventricle may be of the width of a normal infundibulum (Fig. 272), but it may occasionally be grossly dilated (Fig 271).



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ridge), RV, right ventricle, 3V, third ventricle, VS, ventricular septum, VSD, ventricular septal defect

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The diameter, course, and branching of the pulmonary artery and the presence of any valvular stenosis

The over-riding of the aorta and an approximate determination of its degree.

The anatomy of the systemic arteries and their position in relation to the pulmonary artery and its main branches

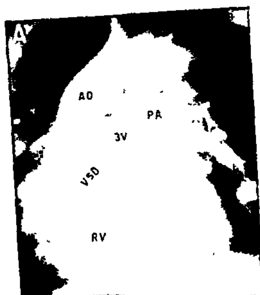
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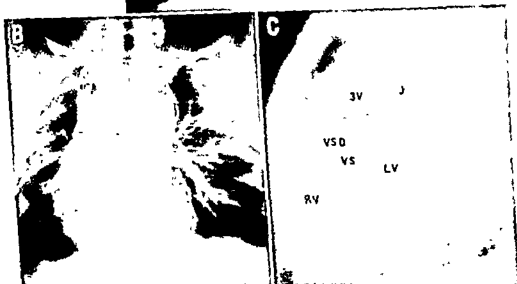


Fig 272 —Tetralogy of Fallot with infundibular stenosis and constriction of pulmonary orifice, rotation, and left superior vena cava Boy, aged 4 (P K. 491010) Infundibular stenosis is confined mainly to ostium infundibuli, which lies just over 2 cm below the valvular plane Third ventricle is large, its width corresponding to that of the normal infundibulum, in frontal projection, it largely overlaps the dorsally displaced pulmonary artery At autopsy, pulmonary orifice was found to be stenosed Only one thickened, malformed cusp is developed, the other is rudimentary Ventricular septum has thick, curved superior border (just above VS in C). Slight dilatation of aorta, which runs in a wide curve Degree of over-riding is difficult to evaluate, owing to rotation [illegible]
wide [illegible]
LV ar[illegible]
septu[illegible]

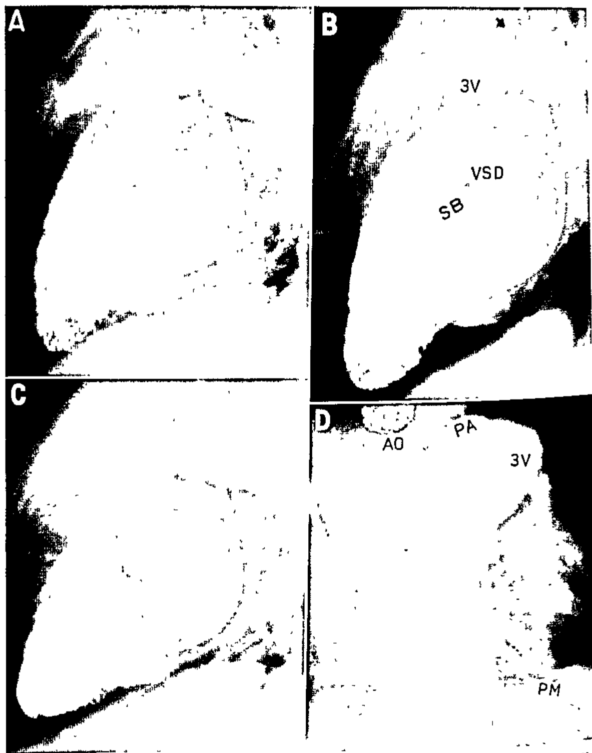


Fig. 271.—Tetralogy of Fallot with infundibular and valvular stenosis Boy, aged 9 (Y G 440424) The infundibular stenosis is confined mainly to the ostium infundibuli *B*, insertion [illegible] of third ventricle Site of [illegible] pulmonary [illegible]), pulmo[illegible] lium passing into left ventricle and aorta, exact site of aortic root is difficult to identify [illegible] hypertrophic trabeculation of right ventricle *AO*, aorta, *PA*, pulmonary artery, *PM*, papillary muscle, *SB*, septal band, *3V*, third ventricle, *VSD*, ventricular septal defect

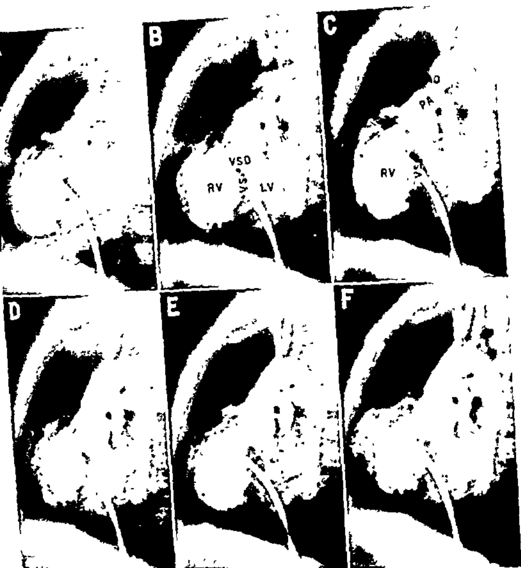

B
C
D
E
F
VSD
RV
LV
VS
RV
PA
VS



Fig. 272 (*cont*)



Fig 273.—Tetralogy of Fallot Girl, aged 1 year (G-B.A 520620) A 50 per cent over-riding of aorta *A–F*, six serial exposures at intervals of 1/12 sec Pulse rate about 170/min Consider- le variations between volume and shape in each picture *AO*, aorta, *I*, infundibulum, *LV* and , left and right ventricles, *PA*, pulmonary artery, *VS*, ventricular septum, *VSD*, ventricular ptal defect (*continued*).

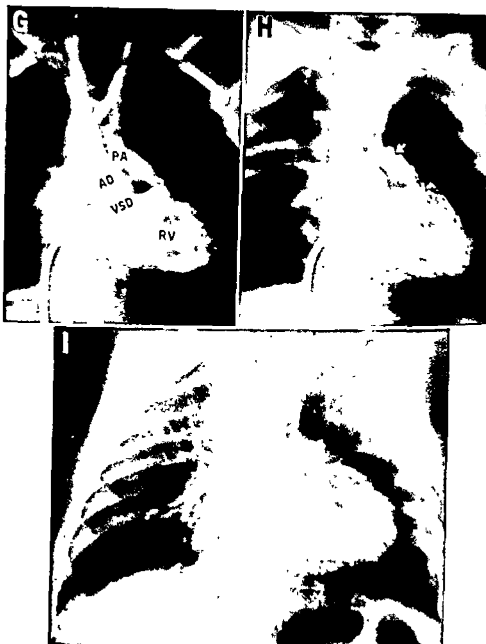


Fig. 273 (*cont*).—*G–H*, marked constriction of whole infundibulum, greatest at ostium infundibuli Arrow in *H* points to semilunar valves. Septum has a broad, curved upper border Defect is somewhat more than half as wide as the slightly dilated, right-sided aorta. Pulmonary artery is short and narrow. *I*, peripherally, extremely scanty vascularity of lungs, circulation is slow. Contrast medium has left the heart and aorta but remains in branches of the pulmonary artery, satisfactory visualization of systemic arteries.

the valvular plane ...
Figs. 269, 273, and 274). In severe

third ventricle, and in some cases ...
complete emptying of the contrast-
blood took place during systole (Figs.
...d 277). Owing to the abnormal inser-
...f the parietal band, retraction of the
...or wall of the ventricle at the level of
...sertion of the band occurs during sys-
... this is best visualized in the lateral
...ction (Figs. 273 and 279) The normal
...acement of the *infundibular region in*
...caudal direction during systole is modi-
... so that there is an inappreciable trac-
... in the anterior and downward direction
... *the contraction* of the apical part of
... ventricle seems to be less marked than
...mally (Fig 273).

The sinus region of the ventricle is usu-
...y large; it is strongly trabeculated and
...rrounded by a thickened wall. As a rule,
... forms the apex

Coincident valvular stenosis was found
...in 24 of the 67 cases with clearly visualized
...enosis or atresia (Figs. 271, 277-280).
...valuation of the valvular region may be
...ifficult, owing to several factors The di-
...mensions of the pulmonary orifice and the
...bulb of the pulmonary artery are often
small, and the orifice is seen in the lateral
view against the contrast-filled aorta. The
caliber of the bulbar segment of the pulmo-
nary artery directly above the pulmonary
ring is *often considerably less than that of*
the distal part of the vessel. This constric-
tion should not be misinterpreted as valvu-
lar stenosis The deformity almost invaria-
bly has a sharp borderline and is most
distinct in systole There is usually continu-
ous dilatation of the lumen in the distal
direction (Figs 272 and 281) Decreased
mobility of the valve may be due to the small
blood flow (Figs 282 and 283) and pro-
vides no definite grounds for a diagnosis of
valvular stenosis We made this diagnosis
only when the valve exhibited the typical
appearance of a domed membrane, which
was usually thickened, protruding into the
...

most easily and ...
infundibulum was dilated only moderately
or not at all (Figs. 281 and 284-286). In
such cases the orifice could be identified
with the same degree of *certainty* as in pul-



Fig 274 —Tetralogy of Fallot with infun-
dibular and valvular stenosis and dextrocardia.
Girl, aged 5 (M P. 481020) Parietal band
causes compression from left side, and septal
defect lies in left superior segment of sinus
region. Small third ventricle. PA, pulmonary
artery, PB, parietal band, RV, right ventricle,
3V, third ventricle, VSD, ventricular septal
defect

monary stenosis with intact ventricular sep-
tum. In other cases, evaluation was made
more difficult by the fact that emptying was
not as constantly in the form of a jet, which
showed the width of the orifice

In many of the cases, the valvular plane
of the pulmonary artery lay far up and to
the back. A characteristic feature is that the
main trunk of the pulmonary artery often
runs in a markedly dorsal direction (Figs.
271, 272, and 283); consequently, in the

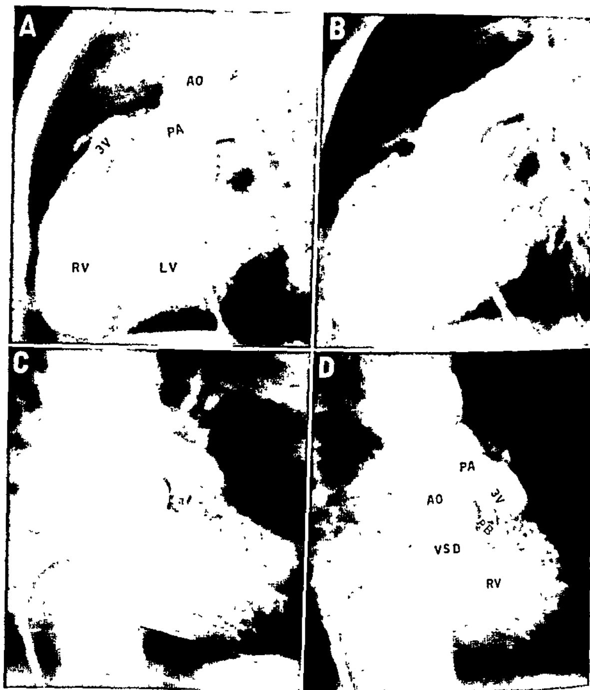


Fig. 275.—Tetralogy of Fallot Boy, aged 3 (G J 500723) Great constriction of whole infundibulum, greatest in ostium infundibuli. Third ventricle is tubular, in frontal projection, it overlaps the dorsally displaced pulmonary artery. Wide aorta is about 50 per cent over-riding Slight decrease in width of pulmonary artery *D*, considerably decreased blood flow in pulmonary circulation *AO*, aorta, *LV* and *RV*, left and right ventricles, *PA*, pulmonary artery, *PB*, parietal band (dextrodorsal conus ridge), *3V*, third ventricle, *VSD*, ventricular septal defect.

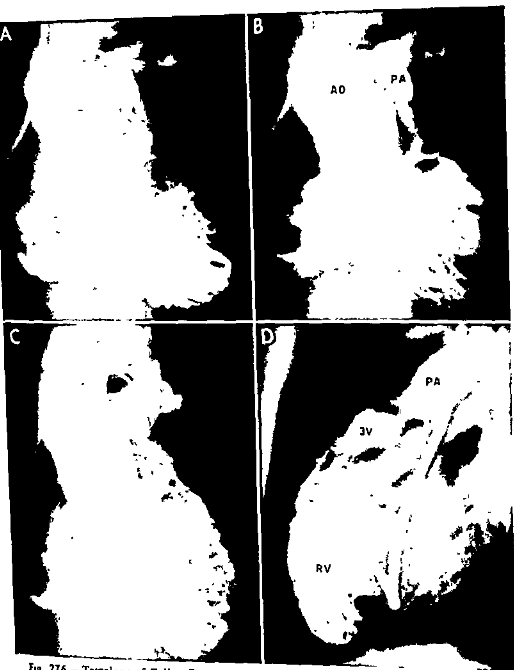


Fig 276 — Tetralogy of Fallot Boy, aged 13 (P K 430730) Marked displacement of both parietal and septal bands, with considerable constriction of the ostium infundibuli in particular, but also of the greater part of the infundibulum *AO*, aorta, *PA*, pulmonary artery, *RV*, right ventricle, *3V*, third ventricle

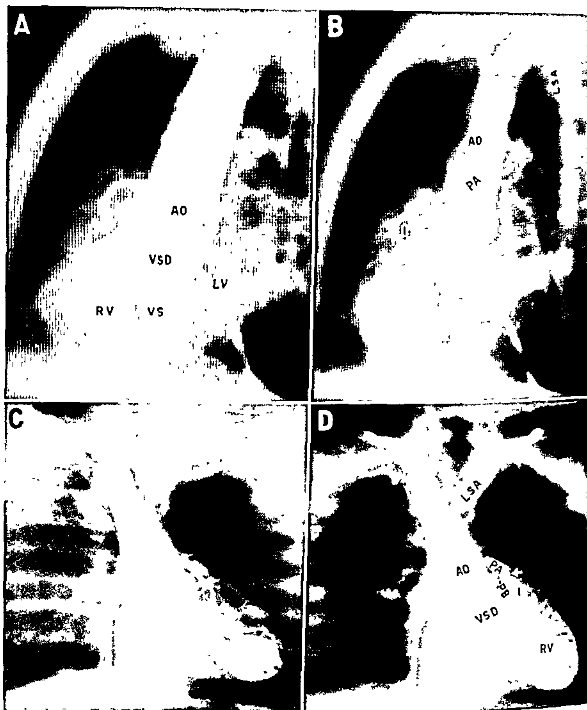


Fig. 277a —Tetralogy of Fallot with infundibular and valvular stenosis Boy, aged 8 months [illegible] of valvular [illegible] nication be- [illegible] aorta, *I*, infundibulum, *LV* and *RV*, left and right ventricles, *PA*, pulmonary artery, *PB*, parietal band (dextrodorsal conus ridge), *VS*, ventricular septum, *VSD*, ventricular septal defect.



Fig 277b —Same case as in Figure 277a Dextrodorsal conus ridge is displaced forward and left and forms an arch over the septal defect (arrow in *A*) Whole infundibulum is constricted [illegible]

[illegible] ventricle Pulmonary artery is narrow and aorta wide. *AO*, aorta, *I*, infundibulum, *LV* and *RV*, left and right ventricles, *MV*, mitral valve, *PA*, pulmonary artery, *PB*, parietal band (dextrodorsal conus ridge), *PM*, papillary muscles, *TV*, tricuspid valve, *VS*, ventricular septum



Fig 278 —Tetralogy of Fallot with inf[illegible] 460207) Cu[illegible] about 50 per[illegible] fundibulum, [illegible]



Fig. 279.—Tetralogy of Fallot with infundibular and valvular stenosis. Boy, aged 7 (
460930). Infundibular stenosis is mainly confined to ostium infundibuli Insertion of pari
band is on anterior wall (ventrally and below *3V* in *C*) Large sinus region, trabeculation hy
B points to fused cusps,
conus ridge), *RV*, right

Fig. 279 (*cont.*)

Fig 280.—Tetral[illegible] dibular stenosis, the [illegible] (A and C) Unusual angulation between pulmonary artery and infundibulum *AO*, aorta, *I*, infundibulum, *PA*, pulmonary artery; *RA*, right atrium, *RV*, right ventricle

frontal view it is depicted with a distinctly axial course. The artery is sometimes curved, with a downward convexity (Figs. 287 and 288). In other cases it has an essentially normal course.

The width of the pulmonary artery varied greatly. It was generally narrow (Figs. 273, 287, 291, and 293) and often short. Poststenotic dilatation (Figs. 279 and 283) was not as common in our series as in those of other authors (659), being present in only 13 cases. The dilatation was not confined to those cases in which there was concurrent or isolated valvular stenosis. In some cases, the bulbous part of the artery was wider than the distal part (Fig. 275). The dilatation sometimes involved the main branches as well (Figs. 254, 272, 284, and 285). It occurred chiefly when valvular stenosis was the dominating impediment to outflow and in stenosis of the origin of the main branches The latter type of stenosis has been described by many authors (e.g. 619). As a rule, the main branches were narrow, as were the vessels in the hilum (Fig 273). The main branches of the artery were usually well filled with contrast medium; *this permitted* the choice of anastomotic operation to be made on the basis of the angiocardiogram. In not a few cases the caliber of one of the branches was so small that an anastomosis to a systemic artery was regarded as out of the question One of the main branches was lacking entirely in three cases (Figs. 290 and 291).

Over-riding of the aorta is best diagnosed in the lateral projection During the injection of contrast medium, pressure increases in the right ventricle, and some of the contrast-mixed blood is expelled through the septal defect into the left ventricle (Figs. 292-295). Vogelpoel *et al.* (676) succeeded in filling the aorta with contrast medium at selective angiocardiography in 11 of 16 cases of tetralogy of Fallot. In our series, the aorta could be demonstrated in all 69 cases. Thus, distinct filling of the aorta was obtained even in those cases in which over-riding of the vessel was minimal or lacking and the septal defect was so small that the systolic pressure in the right ventricle greatly exceeded that in the left. In rapid serial depiction, the pulmonary artery generally is visualized before the aorta. Since the course of the septum is mainly frontal, both the septum and the septal defect are usually well visualized in the lateral projection The defect can,



Fig. 281.—Tetralogy of Fallot. Boy, aged 9 (S.H. 451227). Considerable valvular stenosis and inappreciable constriction of infundibulum. The bulb of the pulmonary artery is small and short. *Stenosis at the origin of the right main branch, with poststenotic dilatation.* Left pulmonary artery hypoplastic *PA*, pulmonary artery, *I*, infundibulum, *RV*, right ventricle, *AO*, aorta





Fig 282 —Tetralogy of Fallot. Boy, aged 8 (J.W. 450227) *A*, short third ventricle, lying immediately below valve Constriction involves both ostium and subvalvular part of the infun-



Fig. 282 (*cont.*)

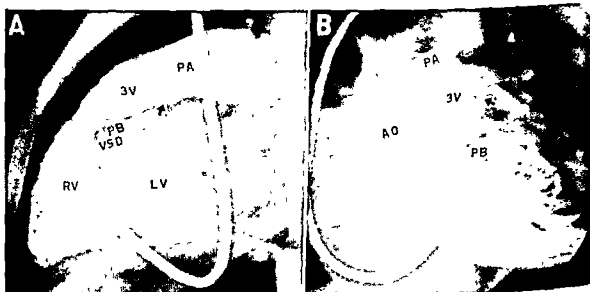


Fig. 283.—Tetralogy of Fallot. Boy, aged 8 (R B. 450903). Stenosis mainly involves the ostium infundibuli. Large third ventricle and slightly dilated pulmonary artery. Incomplete opening of semilunar valves (arrows), presumably owing to decreased blood flow. A number of contrast-filled channels are seen at level of ostium infundibuli *AO*, aorta, *LV* and *RV*, left and right ventricles, *PA*, pulmonary artery, *PB*, parietal band (dextrodorsal conus ridge), *3V*, third ventricle, *VSD*, ventricular septal defect

however, often be seen as well in the frontal view at the beginning of injection, before the overlapping structures have become filled (Fig. 287).

The position of the aortic root in relation to the septum presented considerable variations, it was estimated approximately according to the following criteria:

1 No or inappreciable over-riding: the aortic root was projected in line with or slightly arose entirely from the right ventricle (Figs. 301 and 302).

Demonstration of over-riding of the aorta is based on a study of the anatomy of the outflow tract. It does not take into consideration the quantity of contrast medium that can be demonstrated in the aorta or whether this occurs late or early in the serial depiction.

The demonstration of over riding of the

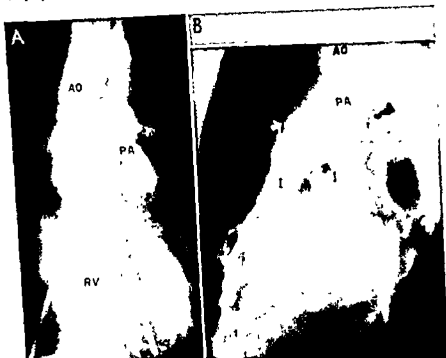


Fig 284 —Tetralogy of Fallot Man, aged 20 (I N 341116). Combination of severe valvular and slight infundibular stenosis No over-riding of aorta The aortic valve lies at the same level as the pulmonary valve. *AO*, aorta, *I*, infundibulum of right and of left ventricle, *PA*, pulmonary artery, *RV*, right ventricle

behind the septal wall of the left ventricle One case in which there was no over-riding was classified as tetralogy of Fallot on the grounds of the clinical features (Figs 284, 285, and 286)

2. Over-riding 25 per cent. the anterior wall of the aorta extended to the septal wall of the right ventricle (Figs 270, 288, 290, and 297)

3 Over-riding 50 per cent the aorta sat astride the septum

aorta is certainly of no apparent importance in planning radical surgery by open methods. Taussig and Bauersfeld (651) have, however, stressed the risk of left ventricular failure and pulmonary edema after an anastomotic operation if the aorta arises mainly from the right ventricle, since the left ventricle is

over-riding of the aorta In this case, the main part of the

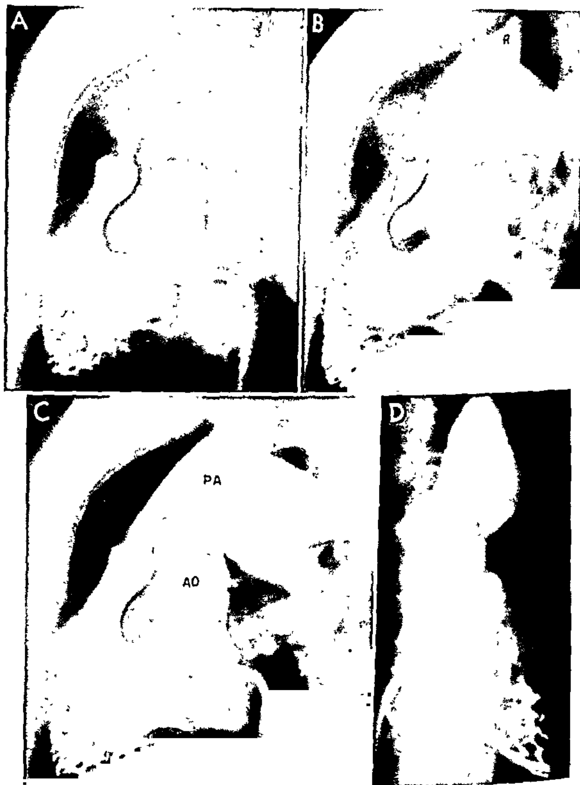


Fig. 285.—Valvular pulmonary stenosis and ventricular septal defect. Boy, aged 14 (S G 401114). A jet of contrast medium is expelled through the 3 mm wide orifice toward the anterior wall of the main trunk. Recesslike bulge (*R*) directly in front of the pericardial reflection Contrast medium passes through the defect in the membranous part of the septum into the left ventricle No over-riding of aorta Right-sided aortic arch Pressure in right ventricle 177/0 mm Hg, in left ventricle 125/0 mm *PA*, pulmonary artery, *AO*, aorta (*continued*).

E F

Fig. 285 (*cont.*)



Fig. 286.—Tetralogy of Fallot. Man, aged 35 (H B 220824). Valvular pulmonary stenosis. Bulb of pulmonary artery short and narrow. Distinct constriction (arrow in *A*) in dorsal aspect of pulmonary artery, at transition between bulb and main trunk. Poststenotic dilatation of distal part of main trunk and whole of main branches *J*, jet, *PA*, pulmonary artery.

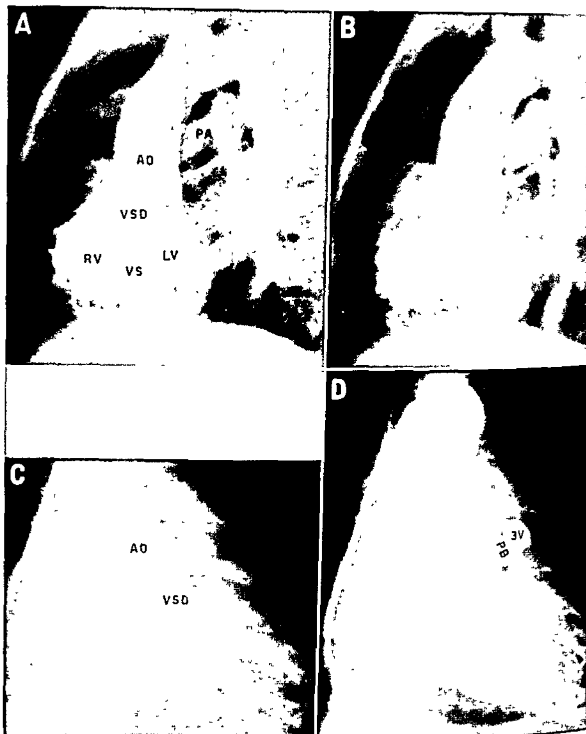
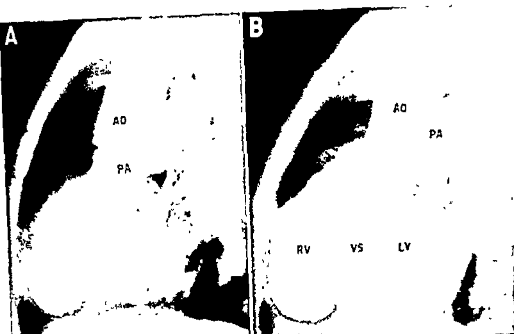


Fig. 287.—Tetralogy of Fallot Boy, aged 5 (K J 480422). Great constriction of whole infundibulum, very small third ventricle, extremely wide aorta, over-riding about 25 per cent, pulmonary artery narrow and runs in a curve, large sinus region, trabeculation hypertrophic *AO*, aorta, *LV* and *RV*, left and right ventricles, *PA*, pulmonary artery, *PB*, parietal band (dextrodorsal conus ridge), *3V*, third ventricle, *VS*, ventricular septum, *VSD*, ventricular septal defect



cles, *PA*, pulmonary artery, *VS*, ventricular septum

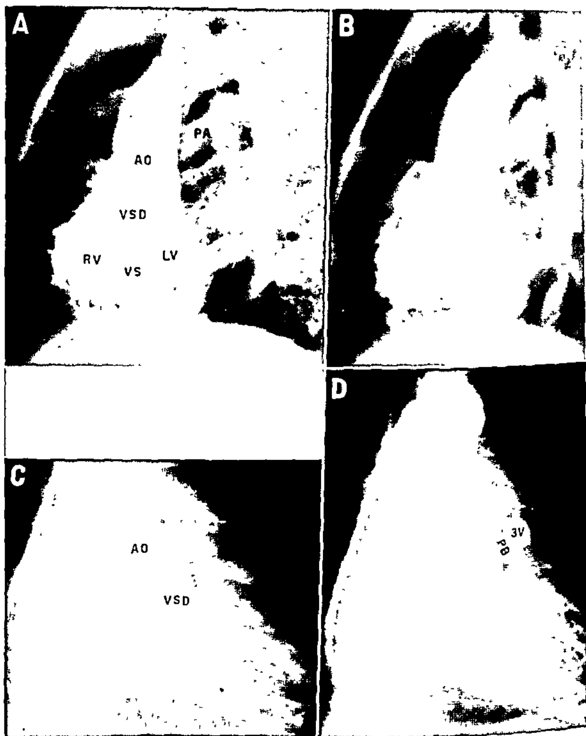


Fig. 287.—Tetralogy of Fallot Boy, aged 5 (K J 480422) Great constriction of whole infu dibulum, very small third ventricle, extremely wide aorta, over-riding about 25 per cent, pulm nary artery narrow and runs in a curve, large sinus region, trabeculation hypertrophic *A* aorta, *LV* and *RV*, left and right ventricles, *PA*, pulmonary artery, *PB*, parietal band (dextr dorsal conus ridge), *3V*, third ventricle, *VS*, ventricular septum, *VSD*, ventricular septal defe



Fig 290.—Tetralogy of Fallot with infundibular and valvular stenosis Girl, aged 5 (M V. 81009) Great constriction of ostium infundibuli (to right of *PB* in C). Small third ventricle Jet spurts from the valvular orifice into pulmonary artery (slightly below *PA* in *A*). Septal defect is almost as wide as the aorta. B, only the left main branch of the pulmonary ... [illegible]

... third ventricle, *VS*, ventricular septum, *VSD*, ventricular septal defect.



Fig. 289.—Tetralogy of Fallot with infundibular and valvular stenosis Boy, aged 5 (L H 470204) Cusps are fused into a dome (arrow in *C*). Large variations in volume in infundibulum (*C* and *D*) Above sinuses of Valsalva (arrows in *A*) main trunk of the pulmonary artery is short and narrow, constriction of both main branches at their origin (slightly to the left of *PA* in *C* and *D*). Wide aorta, over-riding about 50 per cent *AO*, aorta, *I*, infundibulum, *PA*, pulmonary artery, *RV*, right ventricle, *VSD*, ventricular septal defect

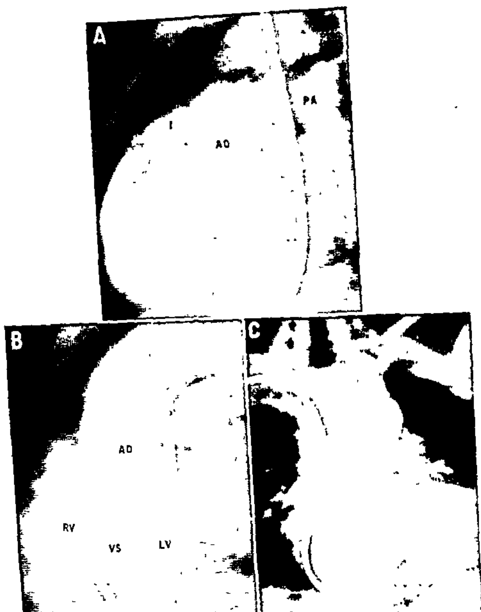
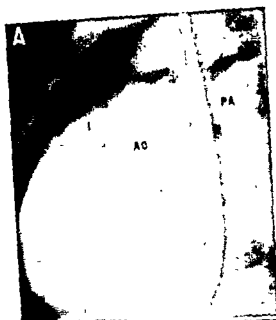


Fig 292.—Tetralogy of Fallot Boy, aged 3 (H T. 490411) Extremely large septal defect. Over-riding of aorta about 50 per cent, and wide communication with left ventricle Aorta is dilated and pulmonary artery is fairly short and narrow. *AO*, aorta, *I*, infundibulum, *LV* and *RV*, left and right ventricles, *PA*, pulmonary artery, *VS*, ventricular septum.



Fig. 291.—Tetralogy of Fallot Girl, aged 5 ... the constricted infundibulum partly overlaps the aortic ... Left branch of the pulmonary artery is narrow, ... about 25 per cent As in all the other cases illustrated, the coronary arteries originate in the aorta *AO*, aorta, *I*, infundibulum, *LPA*, left pulmonary artery, *LV* and *RV*, left and right ventricles, *PM*, papillary muscle *VS*, ventricular septum, *VSD*, ventricular septal defect



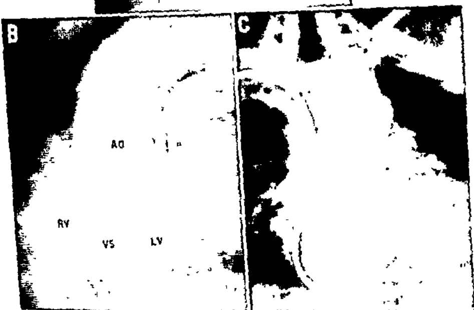
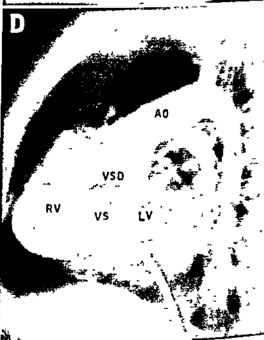


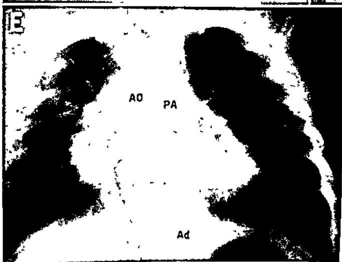
Fig 292.—Tetralogy of Fallot Boy, aged 3 (H T. 490411). Extremely large septal defect Over-riding of aorta about 50 per cent, and wide communication with left ventricle Aorta is dilated and pulmonary artery is fairly short and narrow AO, aorta, I, infundibulum, *LV* and *RV*, left and right ventricles, *PA*, pulmonary artery, *VS*, ventricular septum.











... of Fallot. Girl, ... orta is ... es the right ventricle Communication between left ventricle and aorta is relatively narrow This is presumably the cause of death in left ventricular failure after anastomosis (Blalock's operation) had been performed Both infundibulum and pulmonary artery are narrow *Ad*, descending aorta, *AO*, ascending aorta, *LV* and *RV* left and right ventricles, *PA*, pulmonary artery, *VS*, ventricular septum, *VSD*, ventricular septal defect



Fig 294.—Tetralogy of Fallot. Boy, aged 1 (R S 530812) Infundibular stenosis with great constriction especially of ostium infundibuli (between arrows in *A*) through displacement chiefly of parietal band Third ventricle (3V) fairly small. The ventricular septal defect is clearly visible in *D* *PA*, pulmonary artery, *RV*, right ventricle

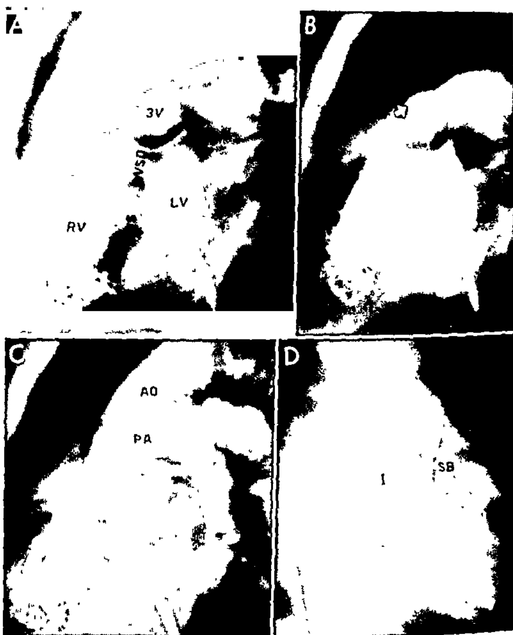


Fig. 295.—*Tetralogy of Fallot.* Girl, aged 1 (K.K. 550918) The outline of the ventricular septal defect (*VSD*) is *distinct* *It* is about 8 mm wide. Great displacement of both parietal and septal bands, with considerable *stenosis* of ostium infundibuli. Valvular pulmonary stenosis (arrow in *B*) as well Aorta 50 per cent over-riding Infundibulum (*I*) of left ventricle wide. *AO*, aorta, *LV* and *RV*, *left* and right ventricles, *PA*, pulmonary artery, *SB*, septal band, *VS*, ventricular septum, *3V*, *third ventricle*



Fig 296.—Tetralogy of Fallot Girl, aged 5 (M L 480614). Fairly marked displacement of both parietal (*PB*) and septal band (*SB*) *with constriction* of ostium infundibuli (*OI*). Numerous small pits and channels are present, particularly on the anterior aspect of the stenosis (arrow in D) Aorta not over-riding or only inappreciably so *AO*, aorta, *CSV*, crista supraventricularis, *PA*, pulmonary artery, *RV*, right ventricle, *3V*, third ventricle (*continued*).



Fig. 296 (*cont.*)

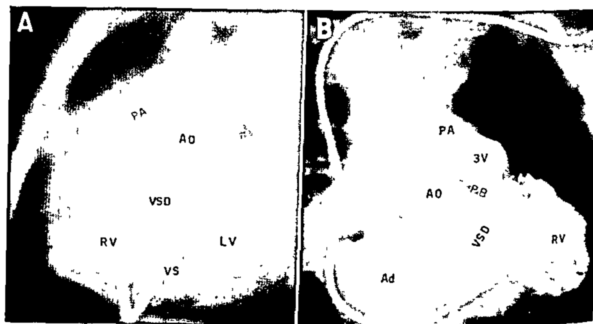


Fig. 297.—Tetralogy of Fallot. Boy, aged 4 (L.P 491023) Marked stenosis, particularly of ostium infundibuli, with small, contrast-filled channels seen at this site Third ventricle is fairly large. Aorta, The sinus region is large and forms the apex. *Ad*, nd *RV*, left and right ventricles, *PA*, pulmonary a ridge), *3V*, third ventricle, *VS*, ventricular septum; *VSD*, ventricular septal defect

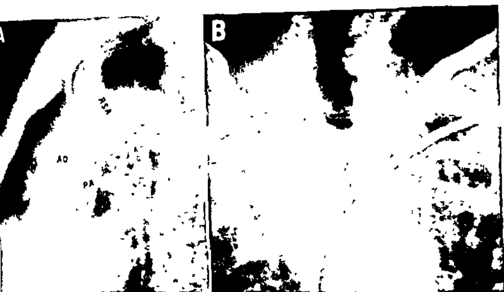
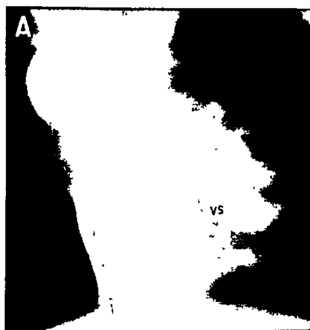
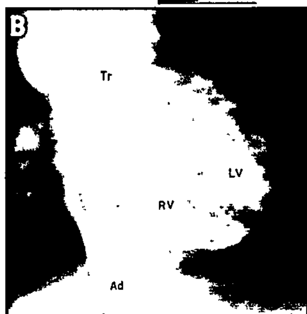
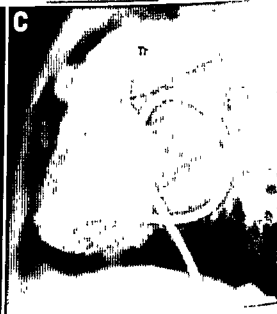


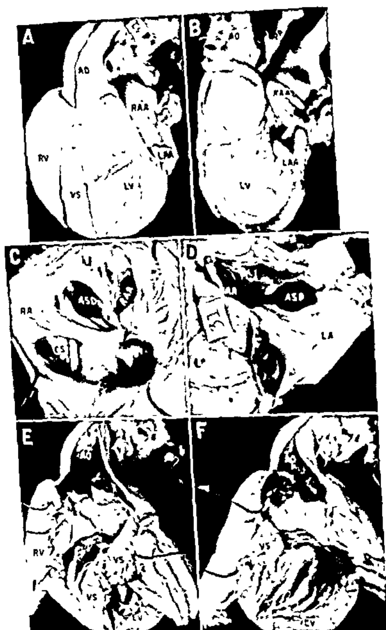
Fig 298 —Tetralogy of Fallot (Blalock's operation). Boy, aged 12 (L K 400930). *A*, right ubclavian artery, which has been brought down and anastomosed with the pulmonary artery, is lled with contrast medium *B*, extensive collateral circulation in superior part of thorax *AO*, orta, *PA*, pulmonary artery, *RSA*, right subclavian artery.







... of infundibulum
common
urse and
observed
r septum



299b.—Same case as in Figure 299a. Both ventricles [illegible]
e [illegible]
[illegible]
[illegible] in E, to left, in F, to right, to show
[illegible] Membranous part of septum is entirely lacking, muscular part ends in a curved margin about 2 cm below the aortic valve. Aorta is extremely wide and [illegible]
[illegible]tum, the parts lying over each ventricle being [illegible]



Fig. 300 —Pseudotruncus and patent ductus arteriosus. Boy, aged 8 (L R. 460604) *A–D*, tip of the catheter, inserted in left ventricle, recoils to the right during injection. *D*, both ventricles … over-riding about 50 per … Large septal defect. Pul- … the ventricles (*DA* in *B*; arrow in *G*). *DA*, ductus arteriosus, *LV* and *RV*, left and right ventricles, *PA*, pulmonary artery, *Tr*, truncus, *VS*, ventricular septum (*continued*)



Fig 300 (cont) —E and ... plane, where pulmonary a ... a vessel runs toward right ... lated bronchial artery



Fig. 300.—Pseudotruncus and patent ductus arteriosus Boy, aged 8 (L.R. 460604). *A-D*, tip of the catheter, inserted in left ventricle, recoils to the right during injection *D*, both ventricles are large, the right is hypertrophic and has a thick wall Wide aorta, over-riding about 50 per cent. Considerable decrease in caliber below orifice of ductus arteriosus Large septal defect. Pulmonary artery is filled from a fairly wide patent ductus and not from the ventricles (*DA* in *B*, arrow in *G*). *DA*, ductus arteriosus, *LV* and *RV*, left and right ventricles, *PA*, pulmonary artery, *Tr*, truncus; *VS*, ventricular septum (*continued*).



Fig. 301 (cont)



Fig 302 —Pseudotruncus Boy, aged 2 months (R.N 530327) No filling of infundibulum of right ventricle nor of pulmonary artery All the contrast medium flows out through a dilated aorta (truncus) The film, exposed during systole, shows the thick ventricular wall The small left ventricle, visible in earlier exposures, empties its entire contents during systole, so cannot be visualized *RV*, right ventricle, *Tr*, truncus



Fig. 301.—Tetralogy of Fallot. Woman, aged 22 (I L 330127). Combined valvular and infundibular stenosis Typical jet in pulmonary artery (*E*) as in valvular pulmonary stenosis The pulmonary artery lies to the left of the aorta Complete transposition of aorta *A–D*, frontal view, *E–G*, lateral view *AO*, aorta, *PA*, pulmonary artery, *3V*, third ventricle (*continued*)





Fig 303b —Functioning anastomosis in tetralogy of Fallot Same case as in Figure 303a. Blalock-Taussig operation was performed, left subclavian artery (*as*) being used Infundibular stenosis, the right-sided aorta is 50 per cent over-riding The left carotid artery is given off as the last branch of the ascending aorta *AO*, aorta; *PA*, pulmonary artery



Fig. 303a.—Tetralogy of Fallot (Blalock's operation) Girl, aged 10 (B L 450406) cf Figu 303b Right-sided aorta The anastomosing left subclavian artery runs behind the esophagus a causes an impression in it (arrow in *C*).



Fig. 305 —Pseudotruncus. Boy, aged 9 (M C 470601). Pulmonary atresia. The aorta takes its origin entirely from the left ventricle. Well-developed collateral circulation (*D–E*). *AO*, aorta, *LV*, left ventricle.

right ventricle and the septal defect was relatively small. After an anastomosis had been carried out, left-sided heart failure occurred and pulmonary edema developed.

We are well aware of the fact that the passage of contrast medium to the left ventricle through the septal defect may be dependent on the artificial conditions which arise, owing to the rise in pressure in the right ventricle during the injection. Consequently, the passage of contrast medium should not be regarded as conclusive evidence of the direction of the shunt. The same reservation applies to corresponding observations when angiocardiography is performed by means of intravenous injection of contrast medium with the patient in the recumbent position. This is because the possibility cannot be ruled out that the heavy, contrast-mixed blood creates an artificial flow through the septal defect, which lies posteriorly.



Fig. 304.—Pseudotruncus Boy, aged 6 (A.V 510904). Extensive collateral circulation A large collateral (arrow) takes its origin from the anterior wall of the aorta (bronchial artery?).

The anatomy of the systemic arteries should be evident from the angiocardiograms. In one of our cases the subclavian artery was lacking on one side, and the blood flow took place through a collateral circulation In another case, this artery was so short and its course was such that it was considered to be unsuitable for anastomosis to the pulmonary artery. A similar analysis of the vascular anatomy has been used as a guide in considering operation according to Potts's method. Postoperative angiocardiography may also be indicated when reoperation is considered. A functioning anastomosis is illustrated in Figures 298 and 303.

Of our seven cases of pulmonary atresia (pseudotruncus), six were investigated by angiocardiography (Figs. 299, 300 and 302). It was fully evident from the examination that atresia was present in and below the pulmonary orifice. In one case, the main trunk of the pulmonary artery and its branches were filled with contrast medium through a wide ductus arteriosus and a bronchial artery running to the right pulmonary branch of the main trunk (Fig 300). A wide collateral vessel was also visible in another case (Fig. 304). In these two cases the aorta was wide and the degree of over-riding about 50 per cent. In the four other cases, no filling of the pulmonary vessels was visible. In one of these cases, a narrow patent ductus was, however, found at autopsy, and the aorta was almost completely transposed (Fig. 301). In one, the aorta took its origin completely from the left ventricle (Fig. 305). In the remaining case, it was difficult to determine the position of the aortic root in relation to the ventricular septum, owing to the atypical course of the latter (Fig. 299).

b) With pulmonary flow via bronchial arteries. Cases of "true" truncus arteriosus without pulmonary arteries are also included in this group Differential diagnosis cannot be established clinically

2 Persistent truncus arteriosus, when one or both of the pulmonary arteries take their origin from the truncus.

In persistent truncus arteriosus without pulmonary artery branch stenosis the pressure in the pulmonary artery is as high as that in the aorta There is both a right to left and a left to right shunt from the ventricles to the truncus. The pulmonary flow is determined by the peripheral resistance in the pulmonary vascular bed. As a rule, it is so low that pulmonary flow is greatly increased *and cyanosis is therefore only mild* *If the resistance increases,* because of changes in the small pulmonary arteries, pulmonary flow may decrease so that considerable cyanosis develops If only one branch of the pulmonary artery is present, there will be a pronounced reduction in pulmonary flow.

We have observed two cases of persistent truncus arteriosus One was a 17-year-old girl who underwent cardiac catheterization at another hospital, only the roentgenologic examination, including angiocardiography, was performed by us. This patient had a *pulmonary artery to one lung only*, she was so cyanotic that tetralogy of Fallot bad been suspected earlier In the second case the history was as follows

Boy, aged 15 months (P N 530307) — Heart disease was detected at 2 months of age, on account of a murmur Physical development was retarded There was slight exertional cyanosis only Dyspnea occurred even on slight exertion The physical findings were characterized by a marked precordial bulge, a systolic thrill, parasternal lift and a harsh systolic murmur with a maximum over the apex The second sound was loudest over the pulmonary area, it was accentuated and strikingly pure

The electrocardiogram in *this* case showed a right ventricular hypertrophy and, in addition tall P waves over the right precordium.

In cardiac catheterization, the catheter passed from the right ventricle into the truncus arteriosus but not into the branches of the pulmonary artery. Systolic pressure was the same in the ventricle and in the truncus. Oxygen saturation of the blood from the truncus was 67 per cent, and its oxygen content was 2.3 volumes per cent higher than that of the right ventricle. The volume of the right to left shunt could not be determined, since the oxygen content of the blood in the left side of the heart was unknown. This is because it cannot be assumed that the pulmonary venous blood is 95 per cent saturated with oxygen in a case with a considerable pulmonary flow, par- [illegible] No [illegible] on [illegible]ults may be obtained in the presence of any large defect in the membranous part of the ventricular septum with a right to left shunt. Gøtzsche (292) succeeded in one case in advancing the catheter from the truncus arteriosus into one branch of the pulmonary artery, and confirmed the diagnosis in this way. In our two cases, the diagnosis could be made only after angiocardiographic examination. The difficulty of establishing the diagnosis on the basis of findings at cardiac catheterization has also been stressed by Anderson *et al.* (14).

ROENTGENOLOGIC EXAMINATION

The roentgen appearance of the heart in true persistent truncus arteriosus is usually the same as in tetralogy of Fallot with pulmonary atresia Rowe and Vlad (570), however, pointed out that the configuration may be less typical if part of the hypertrophied left ventricle forms a segment in the left border, a bulge appearing at that site at which a concavity is seen in other cases. When a bulge is present at the site of the pulmonary artery, it is caused by the left pulmonary artery and usually has a high take-off (17). *If* pulmonary arteries *are given off from the* truncus, the vascularity of the lungs is increased. It is generally stated that the aortic arch is clearly visualized, and the ascending aorta and the aortic arch are often prominent. A right-

10 Persistent Truncus Arteriosus

IF THE truncus conus ridge fails to develop, the truncus arteriosus does not separate into the aorta and the pulmonary artery (see p. 38). A single artery takes its origin from the ventricles. At the same time, there is a defect in the membranous part of the ventricular septum, which is overridden by the truncus. The defect sometimes involves the whole septum, so that a single ventricle is formed. The aortic arch and the main stem of the pulmonary artery, or both branches separately, are given off from the truncus arteriosus. In rare cases, the sixth branchial arch disappears, and the pulmonary branch on one or on both sides is lacking (216) Still more rarely, the right pulmonary artery is given off from the ascending aorta, proximal to the innominate artery, whereas the left branch is given off from the well developed trunk of the pulmonary artery, which takes its origin in the normal way from the right ventricle (145) The anomaly has been ascribed to abnormal septation of the truncus arteriosus (582) The presence or absence of pulmonary arteries is decisive for the clinical syndrome

When both lungs are supplied by pulmonary arteries, their blood volume is plentiful unless the pulmonary artery branches are stenosed at the origin of the truncus The pressure and oxygen content of the pulmonary artery and the aorta are the same.

When pulmonary arteries are lacking, the lungs receive their blood supply through collaterals, mainly through bronchial arteries. As a rule, the blood volume of the lungs is greatly decreased The clinical features are the same as in tetralogy of Fallot with pulmonary atresia and a collateral circulation through bronchial arteries ("pseudotruncus" or "truncus aorticus" (650)). It is true that, embryologically, they are widely divergent malformations, but the differential diagnosis can be made only at autopsy. Cases of truncus arteriosus with absence of the pulmonary arteries have therefore been classified by us as tetralogy of Fallot with pulmonary atresia (see p. 248). In the latter malformation, there is usually a patent ductus arteriosus which supplies the lungs with blood When the sixth aortic arches are lacking, neither the pulmonary arteries nor the ductus arteriosus can exist (216)

By means of angiocardiography, we have been able to show how the contrast medium leaves the heart only through the aorta and, via the patent ductus, fills the main trunk of the pulmonary artery in a retrograde direction down to the atresic orifice (Fig 300). It is sometimes possible in this way to differentiate during life between a true truncus arteriosus and "pseudotruncus" Consequently, we have used the following clinical classification

1 Tetralogy of Fallot with pulmonary atresia ("pseudotruncus," "truncus aorticus")
  a) With pulmonary flow through a patent ductus arteriosus

b) With pulmonary flow via bronchial arteries Cases of "true" truncus arteriosus without pulmonary arteries are also included in this group Differential diagnosis cannot be established clinically.

2 Persistent truncus arteriosus: when one or both of the pulmonary arteries take their origin from the truncus.

In persistent truncus arteriosus without pulmonary artery branch stenosis the pressure in the pulmonary artery is as high as that in the aorta There is both a right to left and a left to right shunt from the ventricles to the truncus. The pulmonary flow is determined by the peripheral resistance in the pulmonary vascular bed. As a rule, it is so low that pulmonary flow is greatly increased and cyanosis is therefore only mild. If the resistance increases, because of changes in the small pulmonary arteries, pulmonary flow may decrease so that considerable cyanosis develops. If only one branch of the pulmonary artery is present, there will be a pronounced reduction in pulmonary flow.

We have observed two cases of persistent truncus arteriosus. One was a 17-year-old girl who underwent cardiac catheterization at another hospital, only the roentgenologic examination, including angiocardiography, was performed by us. This patient had a pulmonary artery to one lung only, she was so cyanotic that tetralogy of Fallot had been suspected earlier In the second case, the history was as follows

Boy, aged 15 months (P N 530307)

The electrocardiogram in this case showed a right ventricular hypertrophy and, in addition, tall P waves over the right precordium.

In cardiac catheterization, the catheter passed from the right ventricle into the truncus arteriosus but not into the branches of the pulmonary artery. Systolic pressure was the same in the ventricle and in the truncus. Oxygen saturation of the blood from the truncus was 67 per cent, and its oxygen content was 2.3 volumes per cent higher than that of the right ventricle. The volume of the right to left shunt could not be determined, since the oxygen content of the blood in the left side of the heart was unknown. This is because it cannot be assumed that the pulmonary venous blood is 95 per cent saturated with oxygen in a case with a considerable pulmonary flow, particularly in a young child (see p. 372). No definite diagnosis could be established on the basis of these findings. The same results may be obtained in the presence of any large defect in the membranous part of the ventricular septum with a right to left shunt. Gøtzsche (292) succeeded in one case in advancing the catheter from the truncus arteriosus into one branch of the pulmonary artery, and confirmed the diagnosis in this way. In our two cases, the diagnosis could be made only after angiocardiographic examination. The difficulty of establishing the diagnosis on the basis of findings at cardiac catheterization has also been stressed by Anderson et al. (14).

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sided aortic arch is common. The diagnosis can be made during life, by means of angiocardiography, if one or both of the pulmonary arteries can be seen to take their origin together with the aorta and the coronary arteries from a common trunk.

Both of our cases (Fig. 306) presented an appearance similar, in most respects, to that of tetralogy of Fallot with pulmonary atresia. The right ventricle was enlarged and hypertrophic, its anterior surface was greatly bulging, the apex was upturned, and a distinct concavity in the cardiac border was visible at the site of the infundibulum The pulmonary artery could not be visualized; the right atrium was enlarged.

The features were nevertheless divergent to some extent in the two cases. In the 1-year-old boy (Fig. 306) the blood volume of the lungs was increased, the left atrium was moderately enlarged, and the aortic arch—which was left-sided, but could not otherwise be judged in detail—was probably narrow. In the 17-year-old patient, the vessels to the right lung were exceedingly sparse, and both the central and the peripheral pulmonary vessels were excessively narrow and scanty. In the left lung, the vessels were dilated centrally but decreased greatly in caliber in the periphery of the hilum. Consequently, they had the appearance characteristic of pulmonary hypertension in the presence of increased resistance. The total blood volume of the lungs was estimated to be reduced, and there was no enlargement of the left atrium. The aortic arch, which was right-sided, was slightly increased in width.

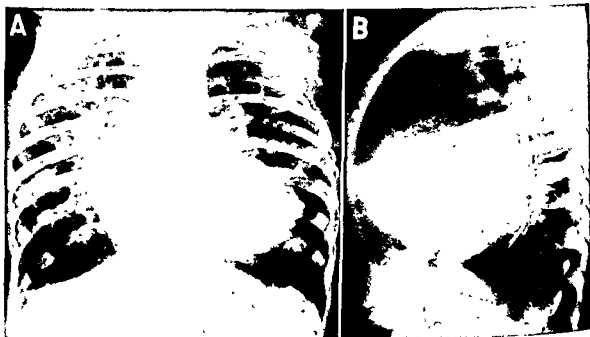


Fig. 306.—*Persistent truncus arteriosus. Boy, aged 1 year (P.N. 530307).* Enlargement of right ventricle, with upturning and angulation of apex as in tetralogy of Fallot; narrow aorta, main trunk of pulmonary artery cannot be identified, greatly increased vascularity of lungs, dilatation of right and left atria

ELECTROKYMOGRAPHY

This examination was performed in one of the cases. The electrokymograms of the pulmonary artery and the aorta are demonstrated in Figure 307. The former had the appearance characteristic of raised pressure in the pulmonary artery. In the latter, which also had an abnormal configuration, the position of the incisura and dicrotic wave farthest down on the descending limb is presumably a manifestation of the increased blood flow and of the raised pulse pressure in the aorta.

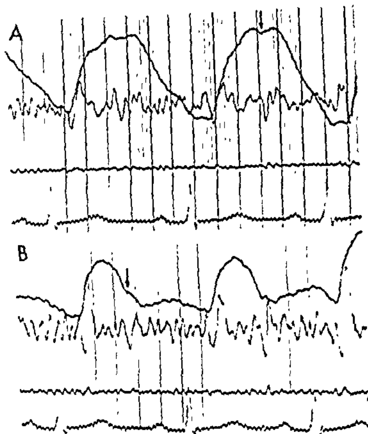


Fig 307.—Electrokymograms in persistent truncus arteriosus Girl, aged 17 (S A 361223). Frequency channels, 10 and 20 cps *A*, left pulmonary artery. Onset of upstroke at normal interval after Q wave; rise continues until end of systole Anacrotic notch is present in its superior part Above it, the rise is slow. Shallow incisura and low dicrotic wave at summit of curve. *B*, aortic arch. Incisura lies far down (arrow) and dicrotic wave is small. Onset of upstroke occurs at normal time.

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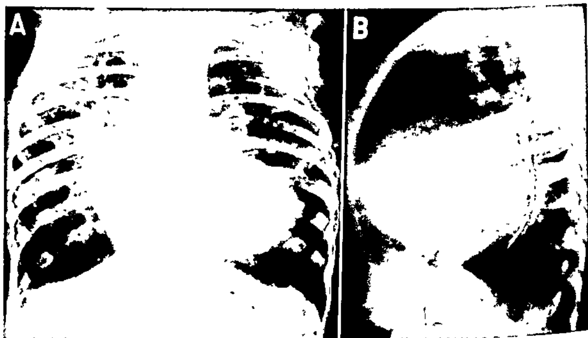


Fig. 306.—Persistent truncus arteriosus Boy, aged 1 year (P.N. 530307). Enlargement of right ventricle, with upturning and angulation of apex as in tetralogy of Fallot; narrow aorta, main trunk of pulmonary artery cannot be identified, greatly increased vascularity of lungs, dilatation of right and left atria.

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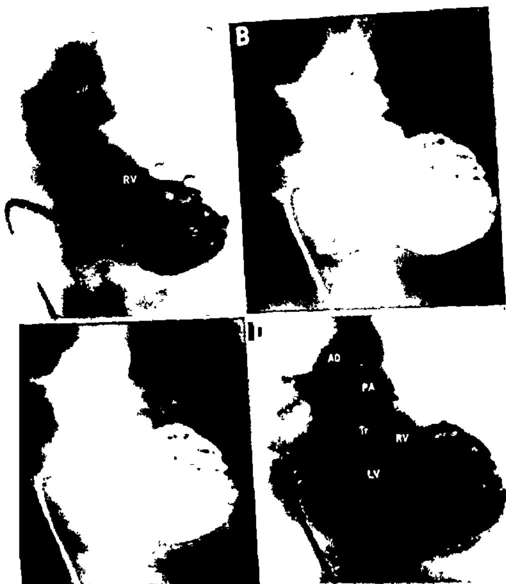


Fig 309.—Persistent truncus arteriosus. Boy, aged 1 year (P.N. 530307) Dilated and hypertrophied right ventricle lies anteriorly to large left ventricle Ventricular septum lies practically in the frontal plane About 50 per cent over-riding of truncus. Pulmonary artery arises from dorsal aspect of truncus, just over 1 cm above the valvular plane, its main branches are of about the same width AO, aorta, *LV* and *RV*, left and right ventricles. *PA*, pulmonary artery. *Tr*, truncus, VS, ventricular septum (*continued*).



Fig. 308.—Persistent truncus arteriosus and atresia of right branch of pulmonary artery Girl, aged 17 (S A 361223) Shape of heart is the same as in Figure 306, with considerable upturning and angulation of apex No dilatation of left atrium or left ventricle. Right ventricle is large and thick-walled. The wide truncus over-rides the ventricular septum by about 50 per cent. Pulmonary artery arises from left side of truncus, about 2 cm cephalad to valvular plane Only left branch of pulmonary artery is visible, it divides into two branches of almost equal width Peripherally, the pulmonary vessels are extremely narrow. No filling of vessels in right lung During injection, the catheter tip slips into the right atrium, and coronary sinus then becomes filled *AO*, aorta, *CS*, coronary sinus, *LPA*, left branch of pulmonary artery, *LV* and *RV*, left and right ventricles, *Tr*, truncus, *VS*, ventricular septum

ANGIOCARDIOGRAPHY

An angiocardiographic examination was made in both cases, the contrast medium being injected into the right ventricle (Figs 308 and 309)

In both cases the wide truncus arteriosus over-rode a large ventricular septal defect. In the 1-year-old, the pulmonary artery arose from the back of the truncus and branched immediately into two wide main trunks The central and peripheral vessels of the lungs were also wide The superior part of the ascending aorta, the aortic arch, and the descending aorta were, on the contrary, narrow.

In the 17-year-old, only the left pulmonary artery had developed. It was wide and arose from the left anterior segment of the truncus Centrally in the left lung the vessels were wide, but they decreased greatly in width in the periphery of the hilum The course of the narrow peripheral vessels was somewhat irregular. The right lung was supplied through fine-caliber bronchial arteries which became filled simultaneously with the intercostal arteries They presumably originated in the descending aorta. The aortic arch, which was [illegible]

[illegible] could a patent ductus arteriosus be identified

In the younger patient, the aortic arch was narrow; the most probable explanation is that the main part of the collected output of the ventricles was shunted to the pulmonary artery, owing to the low resistance in the pulmonary circulation. The flow through the aorta was therefore reduced, and the aortic arch was narrower than normally. This does not, as a rule, occur in tetralogy of Fallot and should therefore be helpful in differential diagnosis.

In the older patient, in whom only one pulmonary artery was demonstrated to be present, resistance in the pulmonary circulation was increased. Consequently, the major part of the blood flowing through the truncus passed into the systemic circuit. As in tetralogy of Fallot, the aorta was somewhat increased in width.

The pulmonary artery and its main branches ran medially in the mediastinum, this is the reason why they could not be identified on ordinary roentgenologic examination, despite their quite appreciable dilatation A similar case was diagnosed at angiocardiography by Abrams and Kaplan (4).

A unilateral pulmonary artery has been observed in conditions other than persistent truncus arteriosus and tetralogy of Fallot. As a rule, the abnormal hemithorax then presents a decreased volume, resulting in retraction of the diaphragmatic dome, a reduction in the breadth of the intercostal spaces and displacement of the mediastinum (718). These features were not present in the second case just described.



Fig. 309 (*cont.*)

ANGIOCARDIOGRAPHY

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11 Ventricular Septal Defect

VENTRICULAR septal defect as an isolated lesion is regarded as one of the most common forms of congenital heart disease. Wood (713) nevertheless pointed out that the incidence figures often given are presumably too high, since other malformations were earlier included under this heading. In combination with other cardiac anomalies, ventricular septal defect is, however, extremely common. In this chapter we shall, however, discuss only the isolated type and such conditions in which, from the functional point of view, the ventricular septal defect is predominant.

Ventricular septal defect and maladie de Roger have sometimes been used as synonymous terms. Taussig (650) distinguishes between maladie de Roger, "a simple perforation in the ventricular wall," and a high ventricular defect, "when the aortic septum fails to meet the ventricular septum." According to her definitions, the defect is small in maladie de Roger and causes only insignificant symptoms. The high septal defects, on the other hand, would give rise to severe symptoms because of their size. As Gøtzsche (292) has stressed, this classification has no historical justification, since Roger did not make such an anatomic distinction. The cases which Roger described had no signs or symptoms, apart from a murmur. Consequently, if the term maladie de Roger is to be used, it is suggested that it be reserved for the mild type.

The clinical features are dependent on the size of the defect. It ranges from a few millimeters to several centimeters in diameter. There are even cases in which the ventricular septum is lacking entirely. Associated malformations are frequently present in such cases.

Descriptions of the anatomy in ventricular septal defect have been given by Rokitansky (567), Selzer (589), and Becu *et al* (49). In most cases, the defect seen from the left ventricle lies immediately below the aortic valve and seen from the right ventricle lies below the crista supraventricularis. Only in about 10 per cent of cases is the defect situated above the crista supraventricularis. It is almost as uncommon to find a defect in the lower part of the septum.

Clinically, the most important and most usual combination is ventricular septal defect and pulmonary stenosis. In cases with a large left to right shunt, a pressure gradient between the pulmonary artery and the right ventricle is a common finding as a result of loss of velocity (see p 120). In other cases, the stenosis is due to malformation of the pulmonary valve or the infundibulum. As we have already emphasized (p. 138), a classification of such combined forms of heart disease is always exceedingly difficult. If there is a left to right shunt and the pressure in the pulmonary artery is normal or raised, the ventricular septal defect seems to be the essential factor, even if the difference between the pressure in the

ulmonary artery and that in the right ventricle is considerable. This is because, in the majority of such cases, the high pressure in the right ventricle is caused by the large septal defect Surgical treatment of the stenosis only is then contraindicated, since it brings about an increase in flow in the pulmonary circulation, whereas the pressure in the right ventricle remains unaltered '

Eisenmenger's complex is characterized anatomically by a subaortic septal defect and an over-riding aorta In contrast to tetralogy of Fallot, there is no stenosis of the infundibulum or the valvular region. The evaluation of the over-riding of the aorta is, however, often difficult even at autopsy. When there is frank over-riding, no problem exists, but difficulties may be encountered in other cases As Selzer (590) has pointed out, an impression is easily obtained of over-riding in all cases of subaortic septal defect, since the shape of the superior part of the ventricular septum is normally spiral.

Lev (424) has stated that "an abnormal architecture of the muscle bands of the right ventricle favors an Eisenmenger complex." In these cases, he found that the parietal part of the crista supraventricularis "may be enlarged, or represented by several muscle bundles with a varied configuration"

Many cases have been classified as Eisenmenger's complex even when the aorta was not over-riding. The diagnosis was then based on the clinical and hemodynamic findings (61, 674) In these cases, there was a right to left shunt through the ventricular septal defect Obviously, the position of the aortic root and the crista supraventricularis is of great importance in the causation of a right to left shunt Such a shunt can, however, occur even with normal aortic root if the pulmonary vascular resistance is high

A small ventricular septal defect constitutes a great resistance to the flow, and a considerable fall in pressure occurs on passage of the blood through the defect The right ventricular pressure may therefore be normal if the defect is small. If a very large defect is present there is, on the contrary, equilibration of the pressure in the two ventricles. Selzer (590) has stated that an intraventricular pressure gradient arises only when the defect is less than half the size of the aortic orifice. If the defect is so large that the ventricles function as a single chamber, the size of the flow in the systemic and pulmonary circulation, respectively, is determined by the total resistance in the two vascular systems If the low resistance in the pulmonary circulation were to persist, the [illegible]

the systemic circulation. With a large ventricular septal defect there is, however, always increased resistance in the pulmonary circulation, it may sometimes even be higher than that in the systemic circulation, so that the shunt is reversed and goes from right to left Not only the size of the defect but also its position may be of hemodynamic importance, this will be discussed later (see p. 371).

The increased resistance is due to anatomic changes in the arterioles (176, 216, 219, 283, 593, 639). The vessel lumen is narrowed by a thick muscular media. There are also thick elastic laminae. Dense collagen is sometimes present in the adventitia This is probably a persistence of the normal fetal type of pulmonary vessels. Later, usually not until after 20 years of age, obliterative intimal lesions appear in the large arteries, whereas the small vessels become dilated (219).

It is conceivable that the pulmonary vascular resistance increases with advancing age The pulmonary hypertension might cause progression of the vascular changes. Exceedingly high resistance resulting in a right to left shunt may, however, occur even in early childhood (74, 700) We have tried, by means of regular roentgenologic examinations, to follow the changes in shape of the pulmonary vessels and the variations in heart volume in 24 children. In the great majority of cases the picture remained fairly unchanged until the age of 5 to 7

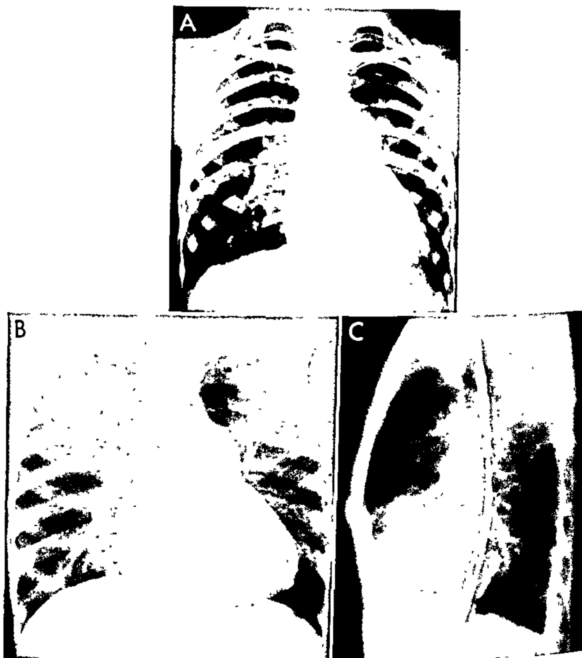


Fig. 310.—Ventricular septal defect and pulmonary hypertension Girl (I.O. 490916) A, at 5 years of age, the pulmonary artery branches are dilated as far as the periphery, but with greatest dilatation centrally B–C, at 8 years, the features are typical of pulmonary hypertension, with narrow, irregularly defined vessels in periphery and wide central vessels No enlargement of left atrium.

years. Thereafter, a reduction in the caliber of the vessels in the periphery was observed in many cases, as well as a decrease in relative heart volume and in dilatation of the left atrium (Figs. 310 and 311) In a few cases the shape of both the pulmonary vessels and the heart, as well as its relative volume, were still unaltered at 10 to 15 years of age (Fig 312) In one case, on the contrary, the typical features of high resistance were present as early as 6 months of age, the picture was unchanged on examination six years later. Thus there is a general tendency to increased resistance with



Fig. 311.—Ventricular septal defect and pulmonary hypertension. Girl (L-L.C. 421209). A–B, at 5 years of age, the width of the vessels is slightly decreased in periphery, but there is still considerable increase in heart volume and enlargement of left atrium C–D, at 13 years, typical picture of increased resistance, with narrow, irregular pulmonary vessels and dilatation of main trunk of pulmonary artery. Inappreciable enlargement of heart, no enlargement of left atrium

rising age, although large individual variations are found Muller *et al* (497) were able to demonstrate, in the dog, the presence of changes in the small pulmonary arteries similar to those in pulmonary hypertension, when a large anastomosis had been made between the aorta and the pulmonary artery This is an indication that increased resistance may arise as a result of high pressure in the pulmonary artery

A left to right interventricular shunt leads to an increased flow through the lungs, the left atrium and the ventricles. Since the shunt from the left to the right ventricle occurs mainly during systole, there is no appreciable increase in the diastolic filling of the right ventricle, but only an increased systolic flow. Consequently, a ventricular septal defect is characterized by a wide pulmonary artery and a normal or narrow aorta, enlargement of the left atrium and left ventricle, but a right atrium of normal size. With pulmonary hypertension, hypertrophy of the right ventricle and

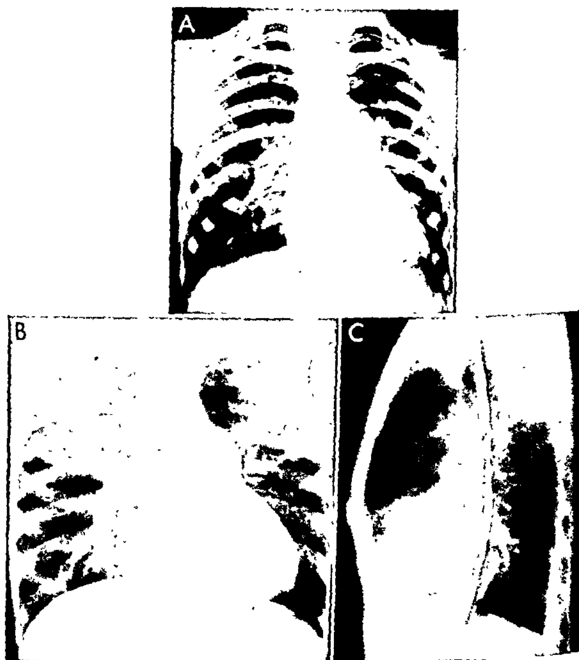


Fig. 310.—Ventricular septal defect and pulmonary hypertension. Girl (I O 490916) A, at 5 years of age, the pulmonary artery branches are dilated as far as the periphery, but with greatest dilatation centrally. *B–C*, at 8 years, the features are typical of pulmonary hypertension, with narrow, irregularly defined vessels in periphery and wide central vessels No enlargement of left atrium.

years. Thereafter, a reduction in the caliber of the vessels in the periphery was observed in many cases, as well as a decrease in relative heart volume and in dilatation of the left atrium (Figs. 310 and 311). In a few cases the shape of both the pulmonary vessels and the heart, as well as its relative volume, were still unaltered at 10 to 15 years of age (Fig. 312) In one case, on the contrary, the typical features of high resistance were present as early as 6 months of age, the picture was unchanged on examination six years later Thus there is a general tendency to increased resistance with



Fig 311.—Ventricular septal defect and pulmonary hypertension. Girl (L-L C. 421209). A–B, at 5 years of age, the width of the vessels is slightly decreased in periphery, but there is still considerable increase in heart volume and enlargement of left atrium. C–D, at 13 years, typical picture of increased resistance, with narrow, irregular pulmonary vessels and dilatation of main trunk of pulmonary artery. Inappreciable enlargement of heart, no enlargement of left atrium.

rising age, although large individual variations are found. Muller *et al* (497) were able to demonstrate, in the dog, the presence of changes in the small pulmonary arteries similar to those in pulmonary hypertension, when a large anastomosis had been made between the aorta and the pulmonary artery This is an indication that increased resistance may arise as a result of high pressure in the pulmonary artery.

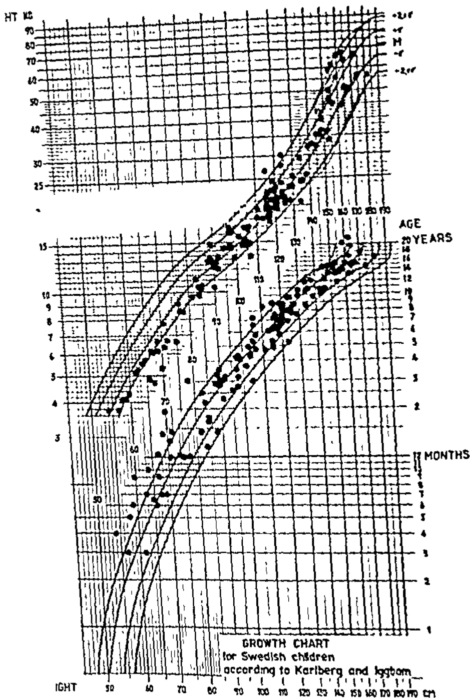
A left to right interventricular shunt leads to an increased flow through the lungs, the left atrium and the ventricles. Since the shunt from the left to the right ventricle occurs mainly during systole, there is no appreciable increase in the diastolic filling of the right ventricle, but only an increased systolic flow. Consequently, a ventricular septal defect is characterized by a wide pulmonary artery and a normal or narrow aorta, enlargement of the left atrium and left ventricle, but a right atrium of normal size. With pulmonary hypertension, hypertrophy of the right ventricle and



Fig. 312.—Ventricular septal defect and pulmonary hypertension Boy (L B. 481023). A, 3 months, large left to right shunt with considerable dilatation of pulmonary vessels far o in periphery. B–C, at 9 years, pulmonary vessels still wide, heart volume considerably i creased, left atrium large

sometimes dilatation of the right atrium are superimposed. In the presence of a very large shunt the systemic blood flow may be decreased.

Our series contains 117 cases of ventricular septal defect 56 boys and 61 girls. The age distribution is shown in Figure 313, in which the physical development is also recorded. Underdevelopment is evident, pa ticularly in the lower age groups Th might be due to the facts that the mo severe cases were investigated at an earl age and that many such patients die early childhood It nevertheless seems mo likely to indicate that improvement tak place as the child grows older.



3 —Physical development in 117 children with ventricular septal defect There is [u]nderdevelopment, particularly in lower age groups (Relevant growth chart was kindly [put at o]ur disposal by Drs P Karlberg and S Iggbom [374, 375])

Our cases were divided into the following groups on the basis of the pressure and the shunts:

	No Pulmonary Stenosis	Pulmonary Stenosis
Group 1 Normal pressure in right ventricle		
a) No demonstrable shunt	20	—
b) Definite left to right shunt: Oxygen content of blood in right ventricle at least 1 volume per cent higher than that of right atrium...	23*	—
Group 2. Systolic pressure in right ventricle above 30 mm Hg but lower than in left ventricle		
Left to right shunt.	27	9
Group 3. Systolic pressure equal in both ventricles		
a) Left to right shunt only	17†	4
b) Mixed shunt, or right to left shunt only.....	16‡	—
c) Complete mixing of blood in both ventricles (functionally, single ventricle)	1	—

*Including 1 case with associated vascular ring and tri- [illegible]

As in all such classifications, it was impossible to adhere to any sharp distinctions. For example, the individual patient may pass from one group into another. This applies in particular to groups 3a and b. If a purely left to right shunt is present at rest, a right to left shunt may develop on exertion. Moreover, it may be difficult to determine whether a decrease in arterial oxygen saturation is caused by a right to left shunt or by incomplete oxygenation of the pulmonary venous blood (see p 372) Nor is there any sharp borderline between groups 2 and 3 Despite such difficulties, this classification—which is essentially in conformity with Selzer's (590)—has proved to be of great value. This is because the various groups correspond to distinct clinical pictures. A description of the anatomy of the malformation in our cases is given together with the account of angiocardiographic examination.

When there is a large left to right shunt and consequently an increased flow through the pulmonary orifice, a systolic pressure gradient between the right ventricle and the pulmonary artery is often found. For, owing to the great velocity of flow, the pressure loss of velocity comes into effect when the pressure is recorded in the main trunk of the pulmonary artery (364) The size of the pressure gradient alone therefore is not decisive for establishing the presence of pulmonary stenosis. The stroke volume must also be taken into account In healthy subjects we have—under basic conditions—found a pressure gradient above 5 mm Hg seldom and above 10 mm Hg never. In the presence of an extremely large left to right shunt there may, on the contrary, be a gradient of up to 20 mm Hg without any anatomic stenosis in the outflow tract of the right ventricle (see p 120). In our series of ventricular septal defect, we had a case with a large shunt and a pressure gradient of 15 mm Hg although no pulmonary stenosis could be demonstrated by angiocardiography. In another case with a gradient of 12 mm Hg but an inappreciable shunt, angiocardiographic examination disclosed valvular stenosis. Thus cases both with and without an anatomic stenosis are found among those with a pressure gradient between 10 and 20 mm Hg. From the practical point of view, there is little value in demonstrating anatomic stenosis of such inappreciable functional importance as that mentioned above. In these cases angiocardiography was not, in fact, performed with a view to demonstrating the presence of stenosis but in order to determine the site of the ventricular septal defect.

Pulmonary stenosis was present in 13 of our cases, it was infundibular in nine and valvular in three. In the remaining case the type of stenosis could not be determined, since angiocardiography was not performed and the withdrawal pressure curve was not decisive. The size of the pressure gradient in both anatomic and "functional" stenosis can be inferred from Figure 344 (p. 370). In anatomic stenosis, the pressure gradient ranged from 12 to 54 mm Hg. A left to right

nt at rest was present in every case. ses with severe stenosis and an interven-ular right to left shunt have been classi-l as pulmonary stenosis (see p. 138).

A ventricular septal defect offers a differ-: resistance to the flow than does the tic orifice, even if the area is the same. ing to the shape of the defect, with its arp margins, the friction loss due to the w through it is greater than that through e aortic orifice. The position of the defect ay also influence the hemodynamics (cf. 371) Kohout *et al* (400) have pointed t that the rapid isometric contraction of e ventricle facilitates outflow to the aorta d that the flow through the defect would therefore start later The fact that the systolic murmur starts immediately after the first sound nevertheless argues against this assumption Provided that diastolic pressure is lower in the pulmonary artery than in the aorta—which applies in the presence of a large left to right shunt—ejection starts in the right ventricle before opening of the aortic valve Consequently, the flow through the defect should even start slightly before opening of the aortic valve

CLINICAL FEATURES

Symptoms are usually late in appearing Great variations are nevertheless found, ranging from patients who live a normal life until an advanced age to those with symptoms even in early childhood. As a rule, this form of heart disease is discovered on a routine examination, owing to the loud systolic murmur. This applied in no less than 105 of our cases, the large majority being detected as early as the first year of life.

Definite symptoms produced by the heart disease before 1 year of age were present in 20 of our cases, all belonging to groups 2 and 3. A poor gain in weight, dyspnea, increased fatigability, and constant respiratory tract infections characterized these patients. An improvement in health can often be noted after the child reaches 1 year of age. In other cases the symptoms become more prominent as the child becomes more active physically. It is, however, striking that some children with an enormously large shunt, pulmonary hypertension and considerable enlargement of the heart seem to be completely unaffected and can take part in strenuous games

Despite the difficulties of making an exact evaluation of the symptoms in children, we have correlated the symptoms to the different types of ventricular septal defect. It can be inferred from Table 7 that such children usually are asymptomatic even if a definite left to right shunt is present If, on the other hand, there is pulmonary hyperte...

... manifestations. Cyanosis at rest was present in six patients One of them had respiratory distress caused by a

TABLE 7 —MOST IMPORTANT SYMPTOMS IN 117 CASES OF VENTRICULAR SEPTAL DEFECT

GROUP*	NO OF CASES	NO SYMPTOMS	FATIGUE	DYSPNEA	CYANOSIS	
					At Rest	At Work
1a	20	18	2	—	—	—
1b	23	19	4	1	1†	—
2	27	20	4	7	—	—
3a	17	6	6	9	1‡	2
3b	16	1	14	15	3	7
3c	1	—	1	1	1‡	—
		With pulmonary stenosis				
2	9	5	1	3		
3a	4	1	2	3		

*For classification see Table 1 page 119
†Associated with vascular ring and tricuspid incompetence
‡Decreased oxygen saturation in pulmonary venous blood

vascular ring; he died a few days after operation for this anomaly, and autopsy disclosed a large ventricular septal defect. Another patient with cyanosis at rest had a large left to right shunt and oxygen unsaturation of the pulmonary venous blood, established by catheterization through a patent foramen ovale. Severe symptoms, as well as signs of heart failure, were present in one case only, it was complicated by auricular flutter and malformation of the tricuspid valve. This patient died at the age of 11 months. Exacerbation subsequently occurred in two cases. The patients became dyspneic at rest but had no edema. Both died during their second year, in connection with a respiratory tract infection. Heart failure does not usually develop until adulthood and is then the most common cause of death. A high mortality in heart failure during the first year of life has, however, been reported by some authors (49, 222, 237, 269, 319, 721) Bacterial endocarditis is a more usual cause of death in children, but did not occur in any of our cases

The following two cases are examples of the mildest and the most severe form, respectively, of isolated ventricular septal defect.

Boy, aged 3 years (J T 500822) —A congenital cardiac defect was discovered during the neonatal period He is not particularly lively, but his mother has never noticed cyanosis or dyspnea even on great exertion. His physical and mental development have been fully normal. A moderately loud, high-pitched systolic murmur can be heard over the precordium, it has a definite maximal intensity over the fourth left interspace (Fig 315, A) Both the electrocardiogram and the roentgenologic appearance of the heart are normal Cardiac catheterization shows normal pressure conditions and no significant left to right shunt The diagnosis of ventricular septal defect was based on the pathognomonic systolic murmur We consider the prognosis to be excellent. His parents have been advised to allow him to lead a normal life

Girl, aged 3 years (E W. 510707) —A congenital cardiac disease was detected at the age of 6 months. She weighed only 2,280 Gm at birth. Her development has been slow, she now weighs 8,500 Gm and did not learn to walk until after she was 2 years old. She is considerably disabled and becomes dyspneic when she runs. Particularly in cold weather, she easily becomes cyanotic when she moves about. Palpation shows a slight precordial bulge and a distinct parasternal lift. A faint systolic murmur can be heard; its maximal intensity is in the fourth interspace to the left of the sternum, and the second sound over the pulmonary area is greatly accentuated The electrocardiogram showed signs of both left and right ventricular hypertrophy Roentgenologic examination disclosed a grossly enlarged heart with a large left atrium and right ventricle and a greatly dilated pulmonary artery The blood volume of the pulmonary circulation was increased. Cardiac catheterization revealed equilibrated pressure in both ventricles and signs of a mixed shunt with decreased arterial oxygen saturation (87 per cent).

In this case, with the great resistance in the pulmonary circulation, the enlargement of the heart and arterial oxygen unsaturation, the prognosis must be regarded as extremely uncertain. Probably, the child can live for another few years, but, unless she can be operated on in the future, it must be presumed that a state of irreversible cardiac failure will gradually develop.

Physical Working Capacity

An exercise tolerance test was performed in only 14 cases (Fig. 314). Patients without pulmonary hypertension were found to have a normal or only slightly decreased working capacity, whereas it was very low in those with pulmonary hypertension If changes in the pulmonary vessels have occurred, the pulmonary flow at work can be increased only by a considerable rise in pressure in the pulmonary artery. But when a large ventricular septal defect is present, the systolic pressure in this vessel cannot exceed the level of the systemic pressure A right to left shunt occurs instead, and these patients often become cyanotic on exertion This implies that the effective stroke volume is small during exercise, which explains the decreased working capacity

Physical Signs

Roger (cited by Brown (107)) gave as the pathognomonic sign a loud systolic murmur with its maximal intensity at the mid-

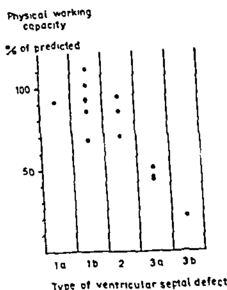


Fig 314 —Physical working capacity, expressed in per cent of predicted value (see p 106), in the different groups of ventricular septal defects For classification, see page 344

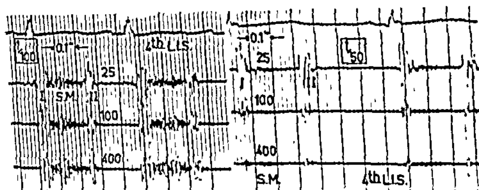


Fig 315 —Phonocardiograms in ventricular septal defect *Left*, boy, aged 3 (J T 500822) Normal pressures no definite shunt demonstrable on cardiac catheterization Systolic murmur has amplitudes up to 400 cps it fills the whole of systole, with maximal intensity in the fourth
left inte
quencie
murmu
high fr
intersp
quency
standa
SM, systolic murmur

dle of the left sternal border, and Brown (107) stated that "little may be added to this original description of Roger." Observations made by a number of authors (107, 202, 360, 503, 554, 620, 713) have nevertheless disclosed that the auscultatory findings in isolated ventricular septal defect are, in fact, subject to considerable variations. For example, a diastolic murmur has been described, and Soulié (620) found no pathologic murmur in five of 70 cases he studied.

We have already stressed the variability of isolated ventricular septal defect. Thus, it ranges from a small, innocent anomaly, with practically no effect on the circulation, to a severe lesion with enlargement of the heart, pulmonary hypertension, and its associated hemodynamic changes. Our material also shows that the physical signs vary greatly in relation to the nature of the malformation. The following description is based on our classification, presented on page 344.

1. The mildest cases (group 1a) exhibited only a high-frequency, blowing systolic murmur, grade 4 to 5, filling the whole of systole. It had a concentrated maximal intensity within a limited area, varying in the different cases from the third to fifth left interspace at the sternal margin. In six of our 20 cases the murmur was sufficiently strong to produce a definite thrill, and in three cases, to produce a slight or suggested thrill. The heart sounds were normal. The phonocardiograms in two cases of this type are shown in Figure 315. In one case (as in one case belonging to group 1b), the murmur was protosystolic. A hypothetical explanation could be that the defect in the muscular part of the septum closes during the muscular contraction in the middle of systole. A murmur of this kind is illustrated in Figure 316.

Neither a precordial bulge, a heaving cardiac impulse, nor a parasternal lift was present in any of the cases in this group.

In group 1b the physical signs were, on the whole, of the same nature as in the preceding group.

Cases with an increased but not equilibrated pressure in the right ventricle have been classified as group 2. In nine of these cases there was coincident pulmonary stenosis. These and four other cases with pulmonary stenosis and equilibrated pressure (group 3a) were characterized by an extremely loud, harsh, high-pitched systolic murmur. They have already been accounted for in Chapter 6 (p. 142, Fig. 133).

Several cases belonging to group 2 had palpatory signs of left ventricular hyper-

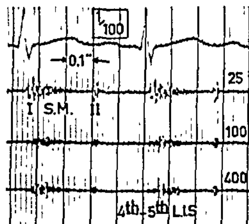


Fig. 316.—Phonocardiogram in ventricular septal defect. Boy, aged 1½ years (T.E. 521008). Small left to right shunt. Oxygen content: of the right atrium 10.2 vol%, of the pulmonary artery 11.7 vol%. Boxed figures denote degree of amplification, other figures denote standard frequencies of the filters. *I*, 1st heart sound, *II*, 2nd heart sound, *L.I.S.* left interspace, *S.M.* systolic murmur.

trophy and a large shunt. A systolic thrill was palpable in every instance. The cardiac impulse was heaving in 11 cases, and a precordial bulge was present in 12. In three cases with the highest right ventricular pressure (57–67 mm Hg) an early systolic pulmonary sound was heard. A third sound was heard in a few cases, otherwise the sounds were normal. The systolic murmur had the same localization and nature as in group 1, but was always of very high intensity (Fig. 317). In nine cases a low-frequency murmur in mid-diastole was heard over the apex. Such a murmur seems to be caused by the large flow into the left ventricle during the phase of rapid filling.

Group 3a contains 17 cases without pulmonary stenosis, there was pulmonary hypertension and a left to right shunt only. The physical signs in these cases consisted of a loud systolic murmur, a definite thrill, a heaving cardiac impulse, a marked or distinct precordial bulge, and, usually, a parasternal lift. In six cases the murmur was protosystolic, with large amplitudes up to the highest frequency range, it was thus identical with that found in most cases of tetralogy of Fallot. The second sound was definitely accentuated, was single or only slightly split, and had a distinct maximal intensity over the second left interspace (470). An example is given in Figure 318, *left*.

When the pulmonary resistance is so great that the shunt through the septal defect becomes mixed, the murmur is extremely faint; in several of our cases it was no louder than the physiologic murmur. Group 3b is composed of 16 such cases. A precordial bulge and parasternal lift were present in all. After the first sound, a more or less marked systolic click was heard, as an expression of both pulmonary hypertension and dilatation. After the faint murmur, systole was ended by a greatly accentuated and not split second sound (Fig. 318, *right*).

It may be inferred from the foregoing description that the physical signs in ventricular septal defect vary according to the anatomy and the hemodynamics. When the pressure in the pulmonary circulation is normal, the murmur lasts for the whole of systole. There is presumably some relation between the size of the shunt and the intensity of the murmur. With a rise in pulmonary pressure, a precordial bulge is added and, finally, a parasternal lift and an

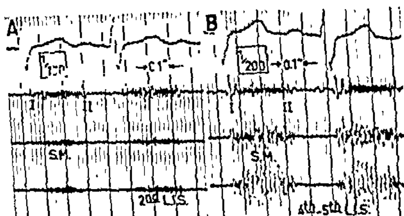


Fig. 317.—Phonocardiogram in ventricular [illegible] left to right shunt and slightly raised [illegible]

early pulmonary sound. A diastolic murmur is common in these cases. With equilibrated pressure and a left to right shunt, the murmur is often protosystolic and the second sound is greatly accentuated over the pulmonary area. When the pulmonary resistance reaches such a degree that the shunt becomes mixed, the murmur is faint. *In these cases, a systolic click and a greatly accentuated pulmonary sound are present.* In cases with pulmonary stenosis load on the left ventricle, but a marked increase in the burden of work of the right ventricle. In such cases, the patients exhibit isolated right ventricular hypertrophy. With a large ventricular septal defect, there are generally both an increased flow and great resistance in the pulmonary circulation. Consequently, the majority of cases present *combined left and right ventricular hypertrophy.*

None of our patients in group 1a exhib-

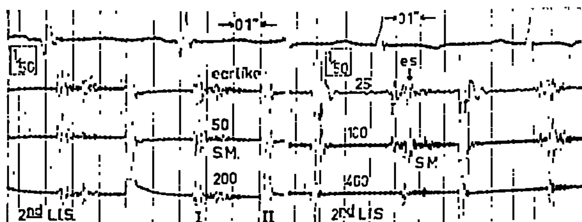


Fig. 318.—Phonocardiograms in ventricular septal defect.

Left, boy, aged 5 (L H. 490114). Equilibrated pressure in both ventricles (80 mm Hg), large left to right shunt, early systolic murmur with maximal intensity over pulmonary area and accentuated 2nd pulmonary sound. Oxygen content of superior vena cava 10 5 vol%; of pulmonary artery 12.6 vol% Boxed figures denote degree of amplification, other figures denote standard frequencies of the filters The elevated isoelectric line in the 200 frequency range is due to amplitudes of the 2nd sound being too high to be managed by the amplifier. *I*, 1st heart sound, *II*, 2nd heart sound, *L I S*, left interspace, *SM*, systolic murmur

Right, girl, aged 12 (L C 421209). Equilibrated pressure in both ventricles (120 mm Hg), mixed shunt, arterial oxygen saturation 90%, very marked early systolic *pulmonary* sound, accentuated 2nd sound over pulmonary area Boxed figures denote degree of amplification, other figures denote standard frequencies of the filters. *I*, 1st heart sound, *II*, 2nd heart sound, *es*, early sytsolic pulmonary sound, *L I S*, left interspace, *SM*, systolic murmur

and a large left to right shunt, the murmur becomes even more intense and fills the whole of systole, with large amplitudes up to the highest frequency range

Electrocardiography

Ventricular septal defect results in an increased burden on both ventricles Their work is determined by the size of the shunt and the total resistance in the pulmonary circulation. If the latter is as high as in the systemic circulation, no shunt takes place and there is therefore no increased

ited signs of ventricular hypertrophy, and left ventricular hypertrophy was present in only six in group 1b. Almost every patient in groups 2 and 3 exhibited either right or left ventricular hypertrophy or both.

In 13 patients an abnormally tall P wave was present in V_1, all of these patients had marked right ventricular hypertrophy. In ventricular septal defect with a large left to right shunt, enlargement of the left atrium can be seen on the roentgenogram, and in some cases electrocardiographic signs of left atrial enlargement can be observed.

One case was complicated by congenital

auricular flutter. A complete right bundle-branch block was found in two cases and an incomplete right bundle-branch block in 12 In 13 cases the P-R interval was prolonged, but a complete A-V block was not present in any case Ventricular septal defect is the malformation that has been regarded to be most commonly associated with congenital, complete A-V block (200). The three cases in our material were, however, combined with other malformations, i e, patent ductus arteriosus (one case), atrial septal (one case), and corrected transposition of the great vessels with pulmonary stenosis and ventricular septal defect (one case) The systolic murmur that is always heard in the presence of a complete A-V block (large stroke volume) has been responsible for an erroneous diagnosis of ventricular septal defect in many cases (140).

ROENTGENOLOGIC EXAMINATION

We made a roentgenologic investigation of 127 patients, 116 of them children Of the 11 adults, all of whom underwent cardiac catheterization, five were sent to our roentgenologic department for consultation.

In isolated ventricular septal defect with a left to right shunt, an increased pulmonary circulation is found on roentgenologic examination It is manifested as a dilatation of the pulmonary artery and its branches, which may exhibit conspicuous pulsations (131, 177, 403, 477)

The findings varied appreciably in relation to the resistance and the blood flow in the pulmonary circulation

When the shunt was small or so slight that it could not be demonstrated on cardiac catheterization (group 1), the roentgenologic appearance was either normal (Figs 319 and 320) or showed slight enlargement of the heart, with or without signs of increased vascularity Marked cardiac enlargement was present in only one case

When there was an increased interventricular flow, with raised but not equilibrated pressure in the pulmonary circulation (group 2), the heart was enlarged and its outline pathologic. The left atrium was dilated and the appendage was prominent to a varying degree, the right ventricle was enlarged and the pulmonary artery and its central branches were dilated. The enlargement of the right ventricle was seen as an increased contiguity of the heart surface to the anterior wall of the thorax. The apex was often curved, its outline was sometimes irregular, and it was distinctly upturned (Figs. 321 and 322). The dilatation of the left ventricle also contributes to the enlargement of the heart, although this is extremely difficult to judge on the roentgenogram, owing to the concurrent enlargement and hypertrophy of the right ventricle A shallow indentation in the cardiac outline above the apex was a common finding (Fig. 321). It indicates displacement of the borderline between the ventricles, but provides no information regarding their relative size. Enlargement of the right atrium was a striking feature in this group The heart volume was sometimes twice the normal volume or more.

The appearance was the same even if the pressure in the pulmonary and the systemic circulation was equilibrated (group 3a), provided the left to right interventricular shunt was large (Fig. 323). When the level of the pressure in the two circulations was the same, but the flow through the pulmonary circulation was smaller, the appearance differed considerably. The peripheral vessels of the lungs were only exceptionally dilated, on the contrary, they were often narrow, whereas the main trunk of the pulmonary artery and its central branches, which exhibited large pulsations, were greatly dilated This is generally interpreted as an expression of increased resistance in the pulmonary circulation. The aorta was of normal width or narrow. The dilatation of the left atrium was not as great as when the blood flow was large and was inappreciable in some cases (Fig 324). All intermediate stages between the two types were observed.

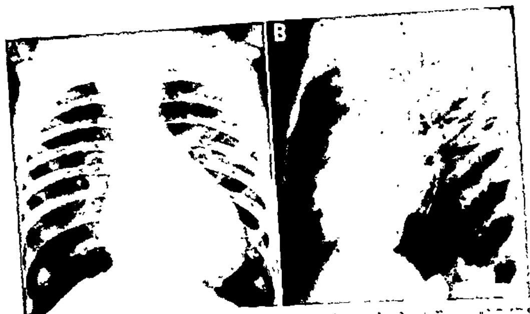
These variations in the roentgenologic



Figs. 319 and 320.—Small ventricular septal defects.

Fig. 319 (*above*) —Girl, aged 6 (S Ö 470127). Appearance of heart is normal and it is not enlarged, normal vascularity of lungs

Fig. 320 (*below*) —Girl, aged 3 (I-L S. 500802) Possibly extremely slight enlargement of left atrium, otherwise no enlargement, normal vascularity of lungs.



dilatation of main trunk of pulmonary artery.

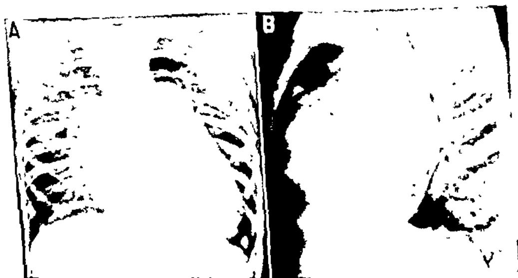
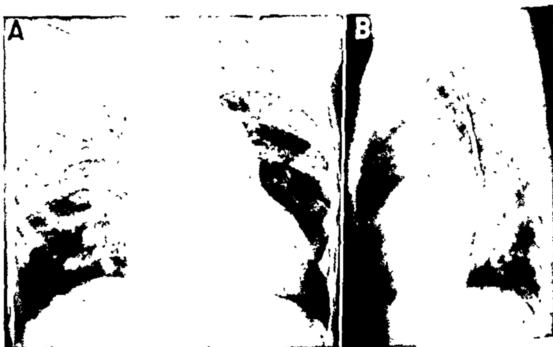


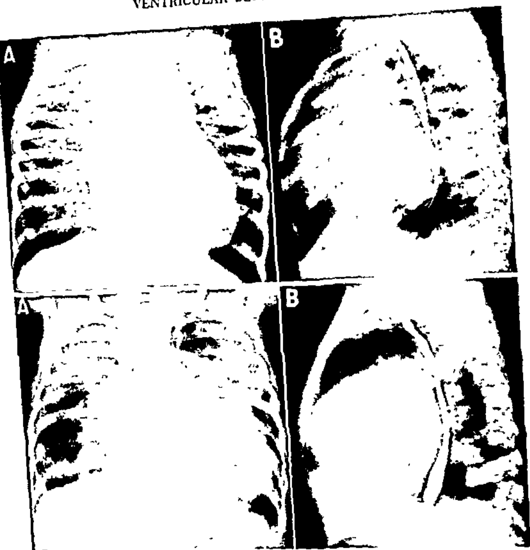
Fig 322 —Ventricular septal defect with large left to right shunt. Boy, aged 2 (A D 500309) Great enlargement of right ventricle, which has hypertrophic shape, considerable dilatation of pulmonary artery and peripheral vessels of lungs, great enlargement of left atrium, narrow aorta



Fig. 323.—Ventricular septal defect with large left to right shunt and equilibrated pressure in ventricles. Girl, aged 4 (A.K. 481015); see Figure 352 (p. 379). Great dilatation of pulmonary artery and peripheral vessels of lungs, very large left atrium.



F. [illegible] small left to right shunt and equilibrated pressure [illegible] ght ventricle with hypertrophic shape, pulmonary [illegible] n central parts of the lungs and normal peripher-



Figs 325 and 326.—Ventricular septal defects with mixed shunt and equilibrated pressure in ventricles

Fig. 325 (*above*) —Girl, aged 3 (E W 510707) Left to right shunt large, enlarged right ventricle, with hypertrophic shape, considerable dilatation of pulmonary artery far out into periphery, left atrium large.

Fig 326 (*below*) —Girl, aged 2 (L-M J 511210), see Figure 347 (p 374) Left to right shunt large, large right ventricle, hypertrophic in shape, pulmonary artery greatly dilated, as are vessels far out in periphery, very large left atrium, cyst formations in right lung.

appearance are clearly illustrated by cases with a mixed shunt (group 3b) In seven small children and in one 13-year-old patient, the dominating feature at both cardiac catheterization and roentgenologic examination was the large flow in the pulmonary circulation (Figs 325-327). In another category, comprising seven patients aged 8 to 37 years and a 2-year-old child, all belonging to the same group, the findings represented a smaller pulmonary flow and increased resistance in th

only three of the 16 cases in group 3b studied by angiocardiography or at autopsy.

In two of them a left to right shunt predominated, and in the other a right to left shunt. The same variations in the roentgenologic appearance and the clinical features were observed in cases with a normal aortic root. Thus, its position does not seem to affect the hemodynamics in the pulmonary circulation in such a way that it is possible, clinically or on the basis of the roentgenologic findings, to say whether or not over-riding of the aorta is present. The roentgenologic appearance is mainly determined by the blood flow and the resistance in the pulmonary circulation.

In ventricular septal defect with a left to right interventricular shunt combined with a local obstruction in the outflow tract of the right ventricle—usually in the infundibulum—the roentgenogram presented the same features as in the uncomplicated cases. We had 12 such cases in our series, an angiocardiographic examination was also made in all but two of them Extensive vascularization of the lungs and great dilatation of the left atrium was found in one of these cases (Fig. 333) and moderate engorgement of the vessels in another In the others, the vascularity was less conspicuous, and in two of them the increase was barely visible (Fig. 334) In this category of patients as well, there was a good correlation between the dilatation of the left atrium and the size of the left to right shunt A small indentation in the cardiac outline marked the site of the stenosis in some of the cases.

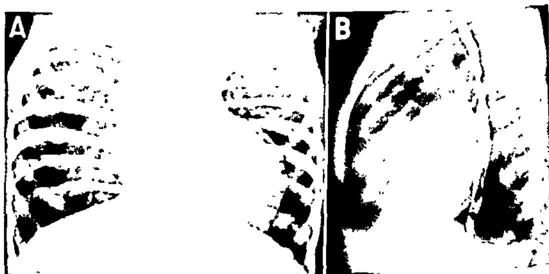
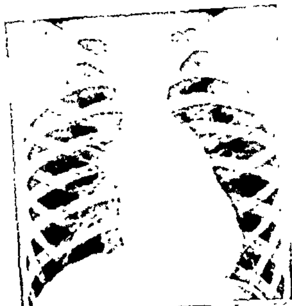


Fig. ... ...ting left to right
shunt ...
385). S...
width of main trunk of pulmonary artery cannot be estimated, considerable increase ... larity of lungs, moderate enlargement of left atrium.

It has been asserted (403) that, in large ventricular septal defects, the dilatation of the pulmonary artery is due to a primary malformation There nevertheless seems to be every indication that the dilatation is essentially dependent on the size of the flow and the pressure in the pulmonary circulation, similarly to the conditions in other congenital anomalies with a left to right shunt We have not infrequently noted an increase in the dilatation of the already widened pulmonary artery and its branches during the first year of life and have interpreted it as an adaptation to the large flow in the pulmonary circulation, as seen in Figure 332

In all forms of congenital malformations of the heart with a left to right shunt, increased vascularity of the lungs is seen on the roentgenogram. In our experience, the combination of this feature with enlargement and, in particular, hypertrophy of the right ventricle, together with dilatation of





Figs 328 and 329 —Ventricular septal defects with small mixed shunt and equilibrated pressure in ventricles

Fig. 328 (*above*) —Girl, aged 9 (E S 440723) Shape of right ventricle hypertrophic, distinct dilatation of main trunk of pulmonary artery, peripherally, reduction in vascularity of lungs, No enlargement of left atrium was seen in the lateral view

Fig 329 (*below*) —Girl, aged 10 (L C 421229) Shape of right ventricle hypertrophic, with curvature of apex, considerable dilatation of main trunk of pulmonary artery, normal or slightly reduced vascularity in periphery of lungs, no enlargement of left atrium



Figs. 330 and 331.—Ventricular septal defects with mixed shunt, large right to left shu during exercise.

Fig. 330 (*above*)—Woman, aged 24 (S B 291103) Hypertrophic shape of right ventric moderate dilatation of pulmonary artery, peripherally, reduction in vascularity of lungs, no largement of left atrium

Fig. 331 (*below*)—Woman, aged 25 (S W. 280821) Hypertrophic shape of right ventric considerable dilatation of pulmonary artery, peripherally, reduction in vascularity of lungs, enlargement of left atrium

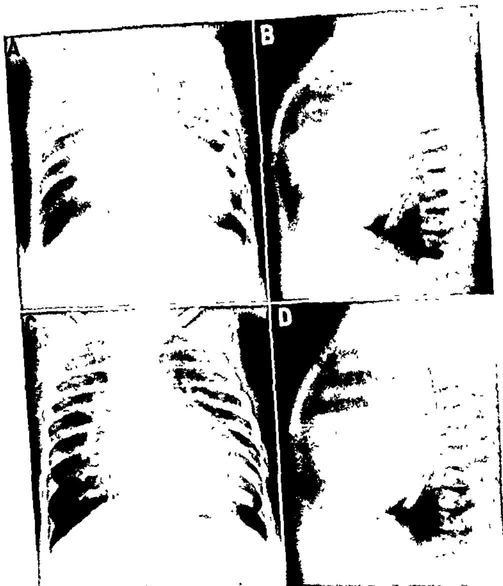


Fig 332 —Roentgenologic progression in large ventricular septal defect with left to right [illegible]

and peripheral vessels of the lungs, shape of right ventricle consistently hypertrophic (*continued*)

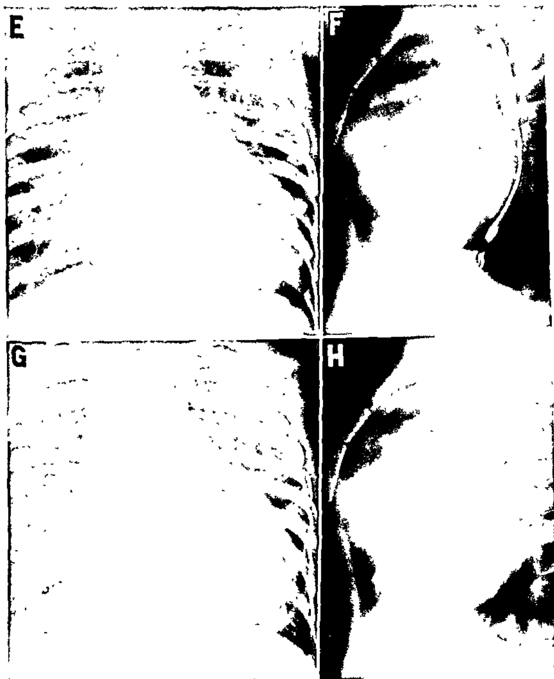


Fig. 332 (*cont.*)

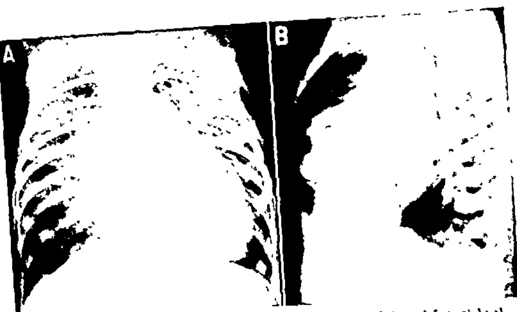




Fig 334 —Ventricular septal defect and infundibular stenosis with left to right shunt Boy, aged 17 (T H 351219), see Figure 370 (p 397) Shape of right ventricle hypertrophic, abnormal insertion of septal band of crista supraventricularis seen ... (arrow), large third ventricle bulges slightly to left (... nary artery, somewhat increased vascularity in periph... left atrium

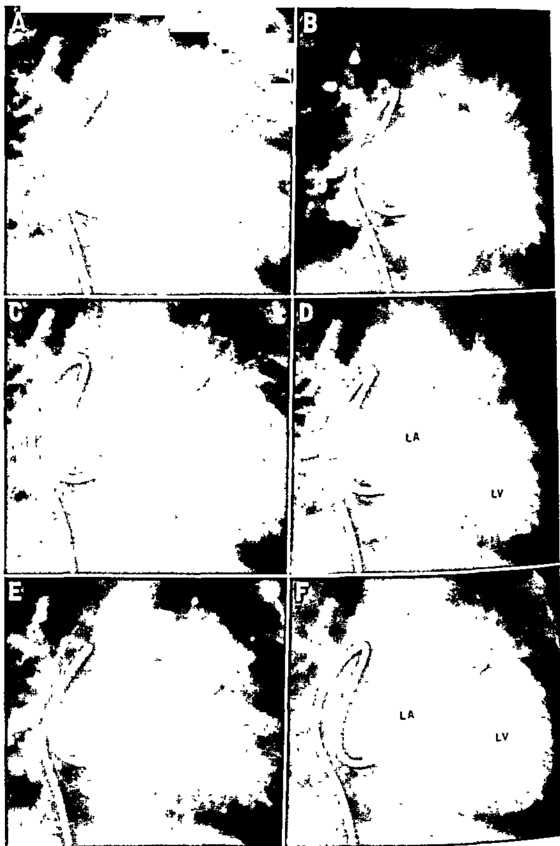


Fig. 335.—Ventricular septal defect with left to right shunt and raised pressure in right ventricle (group 2) Girl, aged 10 months (V U 530527), see Figure 353 (p 380) Great enlarge-



Fig. 335 (*cont*)

ent and incomplete emptying of left atrium (*LA*), dilatation of left ventricle (*LV*) *C* and *J*, late uricular systole, *E*, late ventricular systole

the left atrium, is characteristic of ventricular septal defect with a left to right shunt. Clinically and anatomically, there is enlargement of the left ventricle as well, although it can seldom be demonstrated roentgenologically.

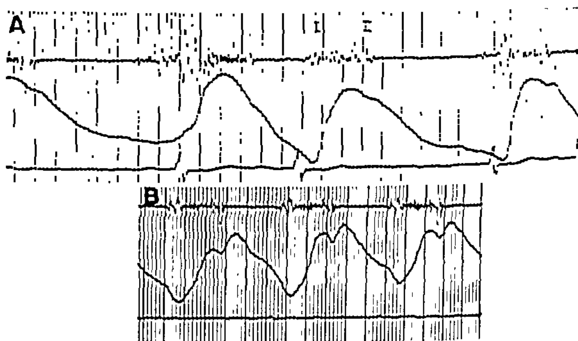
Enlargement of the left atrium is a constant feature which, as far as we are aware, has seldom been stressed in the roentgenologic literature (237, 403, 716). The dilatation may be so great that it causes elevation of the main branches of the pulmonary artery—usually most distinct on the right side—as well as displacement of the main bronchi and an increase in the angle of bifurcation The enlargement of the left atrium can be visualized on angiocardiographic examination (Fig. 335).

The appearance is the same in patent ductus arteriosus complicated by pulmonary hypertension The demonstration of dilatation of the aorta favors this diagnosis.

ELECTROKYMOGRAPHY

This examination was performed in 23 cases. The pulmonary artery tracings were recorded in all, the aortic in the majority and the ventricular and atrial tracings in about half.

In group 1 (cf. p. 344), in which six cases were studied, the appearance of the electrokymograms was normal in every re



...septal defect (grou... (vertex), ... is reduced ... gradient of systolic ascent practically normal The appearance of the tracing varies somewha... with length of the R-R interval. *B*, basal segment of PA. The dicrotic wave is larger and better de... limited from the incisura, which is thereby more prominent than in *A* Upstroke is slower an... its course concave. The incisura is synchronous with the pulmonary component of the 2n... sound ECG unintentionally reduced in amplitude.

spect In group 2, four cases were studied in groups 3a and 3b, two and seven, re... spectively, and, in addition, six cases wit... associated pulmonary stenosis. In groups... and 3, an abnormal pulmonary artery ele... trokymogram was recorded in all but on... case. In this case the pressure in the pu... monary artery was inappreciably raise... (33/6 mm Hg) and the shunt was small

The electrokymogram of the pulmona... artery (Figs 336-340) was characteriz... by a slow, continuous upstroke with i...

creased duration (0.15–0.20 sec), the summit of the curve coinciding with the end of systole. The incisura was high up on the descending limb, and the dicrotic wave was usually diminished. The incisura coincided with the second sound over the pulmonary the tracing were found, but they showed no correlation to the degree of hypertension or the resistance in the pulmonary circulation. The configuration of the tracing is dependent to some extent on the segment of the pulmonary artery over which it is recorded

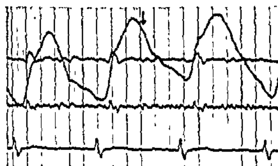


Fig. 337.—Electrok... ...rotic wave are almost obliterated

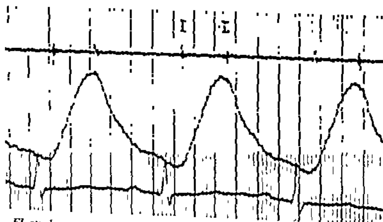


Fig. 338.—El... Girl, aged 12 (I... the end of systc... are obliterated

area Onset of upstroke was sometimes delayed The shape of the curve was rather reminiscent of a sinus wave and is indicative of hypertension in the pulmonary artery In these cases, the lowest systolic pressure in the pulmonary artery was 50 mm Hg, and the highest 125 mm Hg, corresponding to equilibrated pressure in the ventricles Variations in the appearance of (Figs 336, 339, and 340). The shape may also be modified by disturbances of rhythm (Figs 336 and 339).

Similar typical electrokymograms can be recorded in other conditions with pulmonary hypertension, e g , patent ductus arteriosus (p. 553) and primary pulmonary hypertension (p. 825).

The aortic, ventricular and atrial ...

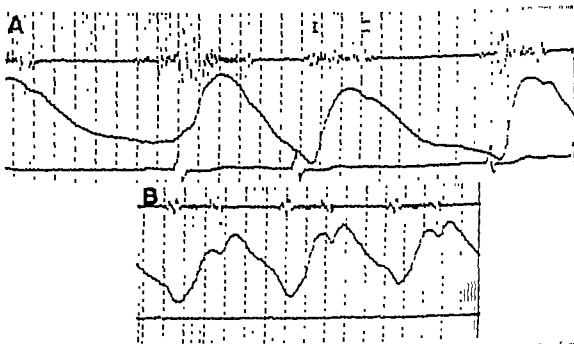
the left atrium, is characteristic of ventricular septal defect with a left to right shunt. Clinically and anatomically, there is enlargement of the left ventricle as well, although it can seldom be demonstrated roentgenologically.

Enlargement of the left atrium is a constant feature which, as far as we are aware, has seldom been stressed in the roentgenologic literature (237, 403, 716). The dila-

ELECTROKYMOGRAPHY

This examination was performed in 28 cases. The pulmonary artery tracings were recorded in all, the aortic in the majority and the ventricular and atrial tracings in about half.

In group 1 (cf. p. 344), in which six cases were studied, the appearance of the electrokymograms was normal in every re-



[illegible] gradient of systolic ascent practically normal. The appearance of the tracing varies [illegible] with length of the R-R interval. *B*, basal segment of *PA* The dicrotic wave is larger and better [illegible] limited from the incisura, which is thereby more prominent than in *A*. Upstroke is slower a[illegible] its course concave. The incisura is synchronous with the pulmonary component of the [illegible] sound ECG unintentionally reduced in amplitude.

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The electrokymogram of the pulmon[illegible] artery (Figs 336-340) was characteri[illegible] by a slow, continuous upstroke with

were particularly conspicuous in the two remaining cases, in which the systolic pressure in the right ventricle below the stenosis exceeded 75 mm Hg The variations were noted to be typical of infundibular stenosis in every respect (Fig. 342).

diagnosed without the help of cardiac catheterization. Even the degree of severity can be judged with fairly great certainty without this examination. The size of the left to right shunt is largely correlated with the increase in heart volume, and the degree of

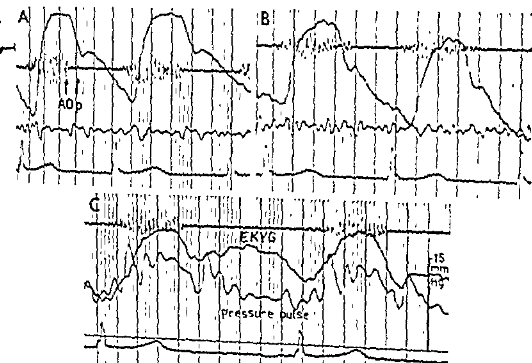


Fig 341.—*Pulmonary* artery electrokymo... shunt through ventricular *septal def*... quency channel around 10 cps *A*, F... Prolonged des... to A Asce... electrokym... pressure cu...

CARDIAC CATHETERIZATION

Cardiac catheterization was performed in all of our cases included in this series A general survey of the results has been given in connection with the classification of ventricular septal defects (p 344) This classification, which is based on the hemodynamic findings, shows good agreement with the symptoms physical findings, EGG, and roentgenologic appearance.

A ventricular septal defect can usually be

right ventricular *strain in the* presence of high pulmonary vascular resistance can be evaluated fairly accurately from the electrocardiograms.

It may, however, be impossible to differentiate cases with very high resistance in the pulmonary circuit (group 3b) from other malformations, *such* as patent ductus arteriosus and atrial septal defect combined with high pulmonary resistance, or from primary pulmonary hypertension *In such* cases, cardiac cathete...

ings had a normal appearance. Electrokymograms of the aorta were not recorded in cases with a narrow aorta.

In nine of the cases studied, there was associated infundibular stenosis. In the four mild cases with a pressure gradient

reminiscent of the tracings in uncomplicated atrial septal defect (p. 435) and in anomalous venous return (p. 515) Common features are a more or less conspicuous plateau-shaped course in systole a delayed predicrotic notch, and reduction of



Fig. 339.—Electrokymogram of pulmonary artery in ventricular septal defect (group 3b Woman, aged 23 (I J. 310201). PCG over pulmonary area Typical pulmonary hypertension curve, appearance varies somewhat with the bigeminal pulse. Decreased amplitude after extra systoles (reduced stroke volume) delayed and more continuous rise in upstroke toward end of systole, incisura synchronous with the 2nd sound *(II)* (1st sound not recorded)



Fig. 340.—Electrokymograms of pulmonary artery (*PA*) in ventricular septal defect (group 3b) Man, aged 37 (E M 170824) PCG over pulmonary area (low and medium frequency ranges) *A*, middle segment of *PA* (vertex), *I*, 1st sound *II*, 2nd sound Slow rise in systole and a plateau at end of systole, incisura and dicrotic wave cannot be identified *B*, basal segment of *PA* Curve is markedly sinus-shaped

across the stenosis of 10 to 30 mm Hg, the tracings had a normal appearance in all essential respects

In three cases in which the pressure gradient ranged from 20 to 40 mm Hg, the pulmonary artery tracings had the configuration illustrated in Figure 341 They were

the dicrotic wave. The typical variations in appearance of curves recorded over different levels of the pulmonary artery are not, on the contrary, electrokymographic features of atrial septal defect.

The aforementioned variations in appearance of the pulmonary artery curves

ere particularly conspicuous in the two remaining cases, in which the systolic pressure in the right ventricle below the stenosis exceeded 75 mm Hg. The variations were noted to be typical of infundibular stenosis in every respect (Fig. 342).

. . . of cardiac catheterization . . . can . . . with- . . . left to right shunt is largely correlated with the increase in heart volume, and the degree of

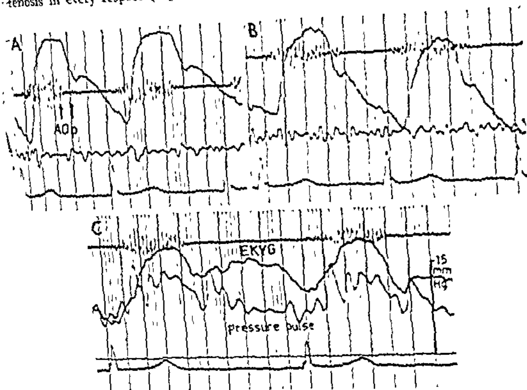


Fig 341.—Pulmonary artery electrokymograms in infundibular stenosis and left to right shunt through ventricular septal defect Boy, aged 11 (I L 430702) PCG over 3rd L I S Fre- . . . electrokymogram (EKYG) Tendency to plateau in both curves, partly masked by vibrations on pressure curve Descending limb and incisura approximately synchronous on the two tracings

CARDIAC CATHETERIZATION

Cardiac catheterization was performed in all of our cases included in this series A general survey of the results has been given in connection with the classification of ventricular septal defects (p 344) This classification, which is based on the hemodynamic findings shows good agreement with the symptoms, physical findings, EGG, and roentgenologic appearance

A ventricular septal defect can usually be right ventricular strain in the presence of high pulmonary vascular resistance can be evaluated fairly accurately from the electrocardiograms.

It may, however, be impossible to differentiate cases with very high resistance in the pulmonary circuit (group 3b) from other malformations, such as patent ductus arteriosus and atrial septal defect combined with high pulmonary resistance, or from primary pulmonary hypertension In such cases, cardiac catheterization is of value

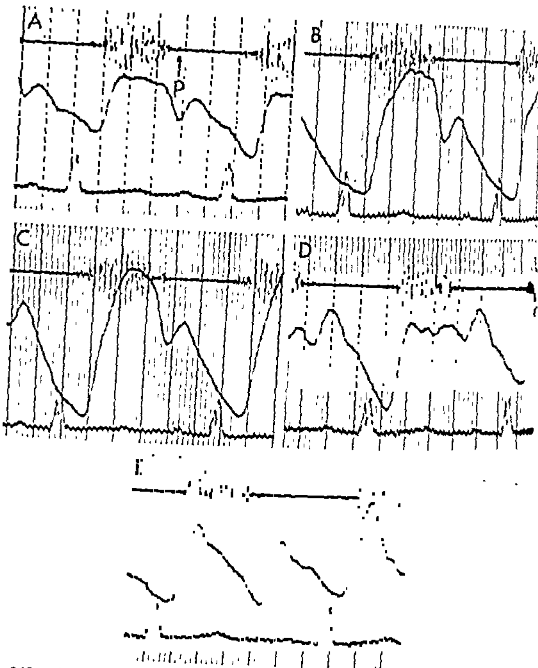


Fig. 342.—Pulmonary artery electrokymograms in infundibular stenosis and left to [illegible] shunt through ventricular septal defect Girl, aged 1[illegible] [illegible]
A, proximal segment of [illegible]
sound. B, somew[illegible]
murmur. Prolon[illegible]
curve D at bord[illegible] [illegible] Almost ordinary pulmonary arte[illegible]
posed [illegible] 0 02 sec earlier. Marked plateau with superi[illegible]
tolic p[illegible] . E, superior part of infundibulum Early sy[illegible]

for the differential diagnosis only if the catheter can be advanced through a ventricular septal defect or a patent ductus. If, on the contrary, the catheter passes from the right atrium to the left, it does not necessarily imply that an atrial septal defect is present. The essential malformation may be a large ventricular septal defect, even if a right to left interatrial shunt can be *demonstrated*, for a ventricular septal defect combined with a marked increase in defect can almost invariably be demonstrated in these cases by angiocardiography with injection of contrast medium into the right ventricle. In such cases with high right ventricular pressure, the position of the defect can generally be identified, and the anatomy of the outflow tract of the right ventricle can be visualized. Malformation of the crista supraventricularis and its muscle bands, as well as the position of the aortic root, can be *established concurrently*.



Fig 343 —Passage of catheter from right ventricle, through a defect in the membranous art of the ventricular septum, via the ascending aorta into the descending aorta Girl, aged 6 M E 470924) Although position of the catheter gives reason to suspect an over-riding aorta, elective angiocardiography showed a normal aortic root

resistance in the pulmonary circuit and severe right ventricular hypertrophy may be associated with a right to left shunt through a patent foramen ovale A systolic pressure in the right ventricle on approximately the same level as that in a systemic artery is an indication of a communication between the ventricles or between the pulmonary artery and aorta Even if such a communication is lacking, the pressure in both ventricles may lie on the same level at rest (cf Pulmonary Stenosis, p 187), but a difference in pressure often appears on great exertion

On the other hand, a ventricular septal Thus, even if cardiac catheterization is not of decisive importance for diagnosing the presence of a ventricular septal defect, it is indispensable for more exact evaluation of the degree of severity of the disease.

On cardiac catheterization, the best evidence of a ventricular septal defect is afforded by passage of the catheter through it, but this is usually successful only when the defect is fairly large In five of our cases, the catheter seemed to pass from the right ventricle directly into the ascending aorta. It is nevertheless difficult to determine on this basis whether or not the aorta is overriding Figure 343 shows passage of the

catheter from the right ventricle, via the defect, into the ascending aorta and aortic arch to the descending aorta. Although the course of the catheter was typical of that in over-riding aorta, angiocardiography disclosed a normal aortic root. In three cases the catheter penetrated the defect and passed into the left ventricle, but could not be advanced into the aorta. In 22 cases the catheter passed through a patent foramen ovale into the left atrium and the pulmonary vein, and in seven of them it continued into the left ventricle

In ventricular septal defect, the oxygen content of the right ventricle should be greater than that of the right atrium A difference of more than 1 volume per cent is usually required for the value to be regarded as significant. The greater the number of blood specimens taken, the smaller the error This increases the possibilities of *demonstrating even a small shunt. It is,* however, often difficult to localize the shunt exactly by means of catheterization Owing to the laminary flow in the atrium and ventricle, no effective mixing of the blood occurs until it reaches the pulmonary artery Several flows with a different oxygen content are present in the right atrium When there is an atrial septal defect, a blood flow from the left atrium with an extremely high oxygen content is superadded. A large proportion of the blood from the atrial septal defect flows directly into the right ventricle

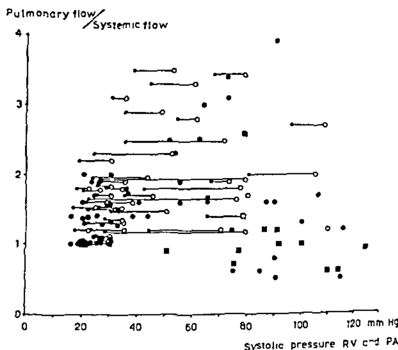


Fig. 344.—Ventricular septal defect relation of size of the left to right shunt (expressed as *the ratio of pulmonary to systemic flow) to systolic pressure in the right* ventricle (open circles) and pulmonary artery (dots) In cases with lower pressure in the pulmonary artery than in the right ventricle, a line denotes the pressure gradient Squares denote cases with a bidirectional shunt. A filled circle or square indicates that there is no pressure gradient

Consequently, if the results of the blood-gas analysis are taken as the only criterion, an atrial septal defect may in some cases be misinterpreted as a ventricular septal defect.

Similarly, in a ventricular septal defect the oxygen content may occasionally be found to be almost the same in the right atrium and ventricle, and higher only when the pulmonary artery is reached, thus simulating a patent ductus It has been assumed *that the flow from the* ventricular septal defect is directed toward the pulmonary orifice This nevertheless presupposes that the

fect lies in the wall between the left ven-icle and the infundibular region of the ght ventricle, which is uncommon. In the eat majority of cases, the flow from the entricular septal defect must pass round ie crista supraventricularis, thus being ixed with the blood in the sinus region of ie right ventricle, before it passes into the ulmonary artery

It has also been stated that a higher oxy-gen content is found if the specimen is aken from the outflow tract. If this denotes he infundibulum, this statement is incor-ect On the other hand, a higher oxygen content is found if the catheter lies on the other side of the crista supraventricularis, immediately adjacent to the septal defect With a normal position of the heart, the ventricular septal defect lies dorsocaudally and to the right of the infundibulum In patent ductus arteriosus with incompe-tence of the pulmonary valve, as well as in aneurysm of the sinuses of Valsalva with a communication into the right ventricle, the gas analysis gives the same results as in ventricular septal defect If a ventricular septal defect is combined with tricuspid incompetence—particularly if the medial cusp is deficient—it may be confused with an atrial septal defect

In most cases of uncomplicated ventric-ular septal defect there is a left to right shunt only, but with extremely high pulmo-nary resistance a right to left shunt may occur. Finally, some cases have a consider-able shunt in both directions This is under-standable in cases with an exceedingly large ventricular septal defect or complete absence of the septum but is more difficult to explain in the presence of a membranous defect with a diameter of about 2 cm It has been assumed that a flow through the defect takes place during diastole as well (103) and that the direction of the shunt differs in systole and in diastole No large flow can, however, occur during diastole, since the diastolic pressure gradient between the ven-tricles is small, particularly in cases with pulmonary hypertension and right ventric-ular hypertrophy In those cases in which we found a large bidirectional shunt, there was a defect between the subaortic region of the left ventricle and the infundibulum of the right ventricle, with malformation of the crista supraventricularis The outflow tracts of the two ventricles then communi-cate, and turbulence during the phase of rapid ejection may produce great mixing of the blood This seems to be a more reason-able explanation of the development of a large bidirectional shunt.

Figure 344 shows the size of the shunt expressed as the ratio of the pulmonary to the systemic flow and its relation to the pressure in the right ventricle and pulmo-nary artery. If the ratio of the pulmonary to the systemic flow is less than 1 there is a predominant right to left shunt or a right to left shunt only. Not included in the figure are two cases with an associated patent ductus arteriosus, one case with an asso-ciated atrial septal defect, and one case with respiratory distress caused by a vascular ring In the last-mentioned case the pulmo-nary artery pressure was normal, but since the oxygen saturation varied considerably at examination, the size of the shunt could not be calculated

The shunt may be considerable without a rise in pressure in the pulmonary artery. In cases with moderate pulmonary hyper-tension, the shunt is also large, but with more severe hypertension there is not a corresponding increase in the shunt. This fact indicates an increase in the resistance in the pulmonary artery. In some of our cases with considerable pulmonary hyper-tension, the left to right shunt was ex-tremely small, owing to high resistance In 12 cases the shunt was mixed, and in five there was a right to left shunt only. In Figure 344, the pressure in both the right ventricle and the pulmonary artery is re-corded, and the respective values are joined by a line; it is thus possible to infer which of the cases are associated with pul-monary stenosis and also its degree. It is seen that the left to right shunt was some-times considerable despite pulmonary ste-nosis

Raised pressure in the pulmonary artery may also be caused by increased pulmonary

catheter from the right ventricle, via the defect, into the ascending aorta and aortic arch to the descending aorta. Although the course of the catheter was typical of that in over-riding aorta, angiocardiography disclosed a normal aortic root. In three cases the catheter penetrated the defect and passed into the left ventricle, but could not be advanced into the aorta. In 22 cases the catheter passed through a patent foramen to the laminary flow in the atrium and ventricle, no effective mixing of the blood occurs until it reaches the pulmonary artery Several flows with a different oxygen content are present in the right atrium. When there is an atrial septal defect, a blood flow from the left atrium with an extremely high oxygen content is superadded. A large proportion of the blood from the atrial septal defect flows directly into the right ventricle.

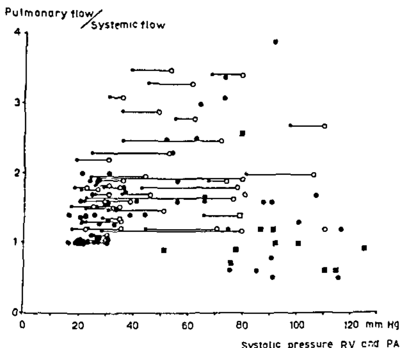


Fig. 344.—Ventricular septal defect. relation of size of the left to right shunt (expressed as the ratio of pulmonary to systemic flow) to systolic pressure in the right ventricle (open circles) and pulmonary artery (dots). In cases with lower pressure in the pulmonary artery than in the right ventricle, a line denotes the pressure gradient. Squares denote cases with a bidirectional shunt. A filled circle or square indicates that there is no pressure gradient

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In ventricular septal defect, the oxygen content of the right ventricle should be greater than that of the right atrium. A difference of more than 1 volume per cent is usually required for the value to be regarded as significant The greater the number of blood specimens taken, the smaller the error. This increases the possibilities of demonstrating even a small shunt It is, however, often difficult to localize the shunt exactly by means of catheterization. Owing

Consequently, if the results of the blood-gas analysis are taken as the only criterion, an atrial septal defect may in some cases be misinterpreted as a ventricular septal defect

Similarly, in a ventricular septal defect, the oxygen content may occasionally be found to be almost the same in the right atrium and ventricle, and higher only when the pulmonary artery is reached, thus simulating a patent ductus It has been assumed that the flow from the ventricular septal defect is directed toward the pulmonary orifice This nevertheless presupposes that the

by only 1 8 volumes per cent, thus indicating a right to left intrapulmonary shunt. The presence of any large arteriovenous anastomoses could be excluded by angiocardiographic examination. All these patients with decreased oxygenation of the pulmonary venous blood were small children, and catheterization was performed under anesthesia This might have led to formation of small atelectases, with resulting variations in the ventilation blood flow ratio throughout the lungs In this event, the decreased oxygen saturation could be explained by a "physiologic" shunting of the blood in the lungs It is nevertheless remarkable that decreased oxygen saturation was present in none of the 32 cases of pulmonary stenosis or tetralogy of Fallot in which the pulmonary venous blood was analyzed, despite the examination having been performed under anesthesia in most cases.

HEMODYNAMICS DURING EXERCISE

The physical working capacity may be normal even in patients with a large left to right shunt (see p 345). A prerequisite is that the effective stroke volume be normal, even with a high pulse rate Case 35/57 in Table 8, belonging to group 1b, had a normal working capacity and a stroke volume which was still normal with a pulse rate of 162 /minute Despite the large flow through the pulmonary circulation during exercise, the pulmonary artery pressure rose only inappreciably In pulmonary hypertension, on the contrary, the working capacity is low, as in Case 39 56, belonging to group 3b (Table 8) The stroke volume was low even at rest An exercise tolerance test could not

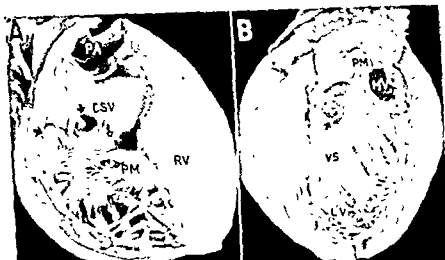


Fig 345.—Large, round defect in membranous part of ventricular septum Defect (right arrow in A) is in upper part of sinus region of the right ventricle, immediately below the right segment of the crista supraventricularis, beside the superior part of the medial cusp of the tricuspid valve In the left ventricle, it lies immediately below the anterior cusp of the aortic valve and is visible in the interior of the infundibulum (arrow in B) Defect is bordered by a small remnant of the membranous part of the septum Right segment of the crista supraventricularis and its parietal band slightly displaced forward Great hypertrophy of trabeculae, slight hypertrophy [illegible] lary muscle, VS, ventricular septum

venous pressure as a result of left ventricular failure. The PCV pressure was, however, *normal in all but six of our cases. In* the exceptions it was only slightly raised, the respective figures being 14, 14, 15, 15, 15, and 20 mm Hg. This rise in pressure is not necessarily an expression only of left ventricular failure, but may be caused partly by the increased flow through the left atrium. In two of the cases in question there was, however, a mixed shunt and only a moderate increase in the pulmonary blood flow. The pressure in the right atrium was normal except in three cases. These cases belonged to group 3, and mean pressure in *the right atrium was 7, 9, and 10 mm Hg,* respectively. Furthermore, the end-diastolic pressure in the right ventricle was raised *only in these cases.*

The possibility of oxygen unsaturation of the pulmonary venous blood in the presence of a large left to right shunt has been discussed. Some authors (59, 283, 589, 593, 674) have expressed the view that oxygenation is normal even with an extremely large flow, whereas others have found it to be decreased in certain cases (518, 714).

Catheterization of the pulmonary vein was performed *in 18 of our cases, and* the catheter then passed through a patent foramen ovale. No demonstrable interatrial shunt was present. In six of these cases the oxygen saturation of the pulmonary vein was decreased (91, 88, 85, 81, 77, and 70 per cent, respectively). All the patients were small children (aged 4 years, 14, 10, 9, 4, and 3 months, respectively), with a large left to right interventricular shunt and pulmonary hypertension In the remaining 12 cases the oxygen saturation of the pulmonary venous blood was 93 per cent *(two cases), 94 per cent (one case),* and over 95 per cent (nine cases). The age of the patients ranged from 6 months to 15 *years, and* most of them had a large left to right shunt.

In two of the cases with decreased oxygenation of the pulmonary venous blood, the oxygen content was also determined during breathing of 100 per cent oxygen.

TABLE 8.—HEMODYNAMICS IN 2 PATIENTS WITH VENTRICULAR SEPTAL DEFECT (In one patient studied both at rest and during exercise, the pulmonary artery pressure was normal, in the other, pulmonary hypertension was present, and no exercise test was performed.)*

Case No	Sex	Age, Years	Body Surface Area, M²	Physical Working Capacity, % of Predicted	Work Load, kg/min	O₂ Uptake, ml/min	Pulse Rate, beats/min	Pulmonary Circulation: AV O₂ Diff., ml/L	Pulmonary Circulation: Cardiac Output, L/min	Pulmonary Circulation: Stroke Vol., ml	Systemic Circulation: AV O₂ Diff., ml/L	Systemic Circulation: Cardiac Output, L/min	Systemic Circulation: Stroke Vol., ml	O₂ Cap., vol %	Art O₂ Sat., %	Shunt, L/min, L → R	Pressure, mm Hg: RA Mean	RV Syst.	PA Syst.	PA Diast.	PA Mean	Br. A. Syst.	Br. A. Diast.	Br. A. Mean
35/57	M	20	2.06	92	Rest	330	82	19.8	16.7	204	25.0	13.2	161	18.8	96	3.5	3	27	21	11	17	141	94	108
					300	915	105	40.3	22.7	216	62.7	14.7	140	19.0	96	8.0		—	32	11	23	142	86	107
					600	1452	130	49.6	29.3	225	84.0	17.3	133	18.6	95	12.0		36	27	15	20	141	77	100
					900	2144	162	62.2	34.6	214	95.9	22.4	138	19.3	95	12.2		49	30	13	21	146	82	101
39/56	M	43	1.78	44	Rest	263	104	102.1	2.6	25	85.2	3.1	30	30.5	92	R → L† 0.5	4	133	133			119	75	92

*For abbreviations see Table 1, p 110.

†Pulmonary venous blood assumed to be 97 per cent saturated.

he analyses were made with the Van Slyke apparatus. Full oxygen saturation was not obtained and the oxygen content increased by only 1.8 volumes per cent, thus indicating a right to left intrapulmonary shunt. The presence of any large arteriovenous anastomoses could be excluded by angiocardiographic examination. All these patients with decreased oxygenation of the pulmonary venous blood were small children, and catheterization was performed under anesthesia. This might have led to formation of small atelectases, with resulting variations in the ventilation blood flow ratio throughout the lungs. In this event, the decreased oxygen saturation could be explained by a "physiologic" shunting of the blood in the lungs. It is nevertheless remarkable that decreased oxygen saturation was present in none of the 32 cases of pulmonary stenosis or tetralogy of Fallot in which the pulmonary venous blood was analyzed, despite the examination having been performed under anesthesia in most cases.

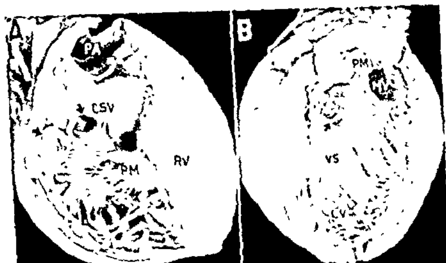


Fig. 345.—Large, round defect in membranous part of ventricular septum. Defect (right arrow in A) is in upper part of sinus region of the right ventricle, immediately below the right segment of the crista supraventricularis, beside the superior part of the medial ... cuspid valve. In the ... and ... nar ... us ... of ... th ... lar ... lar

HEMODYNAMICS DURING EXERCISE

The physical working capacity may be normal even in patients with a large left to right shunt (see p. 345). A prerequisite is that the effective stroke volume be normal, even with a high pulse rate. Case 35/57 in Table 8, belonging to group 1b, had a normal working capacity and a stroke volume which was still normal with a pulse rate of 162/minute. Despite the large flow through the pulmonary circulation during exercise, the pulmonary artery pressure rose only inappreciably. In pulmonary hypertension, on the contrary, the working capacity is low, as in Case 39/56, belonging to group 3b (Table 8). The stroke volume was low even at rest. An exercise tolerance test could not

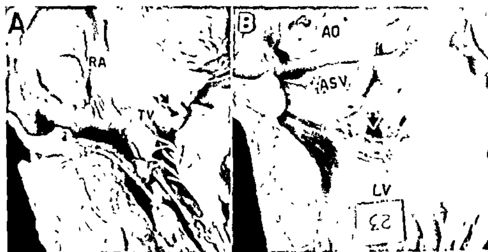


Fig. 346.—Small, fairly round defect in membranous part of ventricular septum (arrow in *B*). Medial cusp of the tricuspid valve is rudimentary and fused to the septum. It covers the septal defect entirely and is perforated by a small hole directly opposite the defect (arrow in A). The chordae tendineae of this cusp are lacking. Appearance of the other cusps is normal. *AO*, aorta; *ASV*, aortic semilunar valves; *LV*, left ventricle; *RA*, right atrium; *TV*, tricuspid valve.

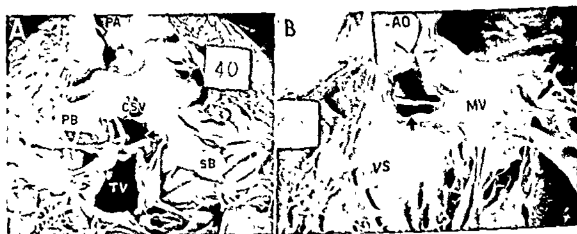


Fig. 347.—Same case as in Figure 326. Large, oval defect in membranous part of the ventricular septum, directly above the medial cusp of the tricuspid valve (arrow) In B, one of the ... segment of the crista supraventricularis ... lso foreshortened and inserted cephalad ... Left anterior cusp of the aortic valve ... is with both bands and trabeculae are hypertrophied. Aorta is not over-riding *AO*, aorta, *CSV*, crista supraventricularis; *MV*, mitral valve; *PA*, pulmonary artery; *PB*, parietal band, *SB*, septal band, *TV*, tricuspid valve, *VS*, ventricular septum

be made during catheterization, but there is reason to assume that the right to left shunt would have increased during exercise.

ANGIOCARDIOGRAPHY

MORPHOLOGIC BACKGROUND —The membranous septal defects are invariably situated in the posterosuperior part of the right ventricle, immediately below the crista supraventricularis, beside and above the medial cusp of the tricuspid valve (Fig 345) They vary considerably in size and are usually round or oval (Figs 345–347). In the left ventricle, they are always found high up under the aortic valve. When there





show

SB, septal band, *TV*, tricuspid valve

is coincident over-riding of the aorta, the septal defect is large and often oval While most of the ventricular defects are related to the membranous portion of the septum, it is exceptional for a defect to be restricted to this portion, as Becu et al (49) have pointed out

The right segment of the crista supraventricularis and its parietal band usually run in an abnormal direction, that is, forward (Fig 348) The parietal band is foreshortened and is inserted more cephalad and farther to the left than normally. There is generally only inappreciable narrowing of the ostium infundibuli.

The defects in the muscular part of the

septum vary greatly in site, shape, and size. In the right ventricle, they are almost invariably situated caudal to the crista supraventricularis and farther to the left of the tricuspid valve (Fig. 349).

When there is an anomaly of the dextrodorsal conus ridge (*see* Chapter 1, on Embryology), a large defect is found in the right dorsal segment of the infundibulum of the right ventricle, and a wide communication between the ventricles (Figs 48, p 46, and 350)

Angiocardiographic examination — The indications for angiocardiographic examination in ventricular septal defect with a left to right or mixed shunt have become successively broadened The constant advances in heart surgery have now made it possible to carry out successful repair of these defects, their demonstration on angiocardiography has therefore become a topical matter It may be presumed that determination of the site of the defect and, if possible, of its size are of paramount importance for consideration of the surgical aspects of the case

From the technical viewpoint, injection of the contrast medium into the left ventricle should be helpful in a study of ventricular septal defect. This is because advantage is taken of the direction of the blood flow in the defect, and optimal density in and adjacent to it can be anticipated. Injection of the contrast medium by this method was made in one of our cases The catheter was introduced into the left ventricle, after it had passed into the left atrium through a patent foramen ovale. The findings are illustrated in Figure 351a The septal defect allowed passage of a wide flow of contrast medium to the right ventricle, it was most clearly visualized during initial filling of the ventricle. The defect was subaortic, and its width estimated as slightly more than half the transverse section of the aorta. The aorta was over-riding The degree of over-riding was difficult to establish, since oblique projections were used. At autopsy one year later, the observations were confirmed in all essentials (Fig. 351b)

Injection of the contrast medium into the left atrium is, conversely, associated with considerable drawbacks The atrium is di-



hypertrophy of all muscular elements of the right ventricle and trabeculae of the left (pressure in the ventricles was equilibrated). The crista supraventricularis and its bands are at the normal site *CSV*, crista supraventricularis, *MV*, mitral valve, *PA*, pulmonary artery; *PB*, parietal band, *SB*, septal band, *TV*, tricuspid valve, *VS*, ventricular septum.

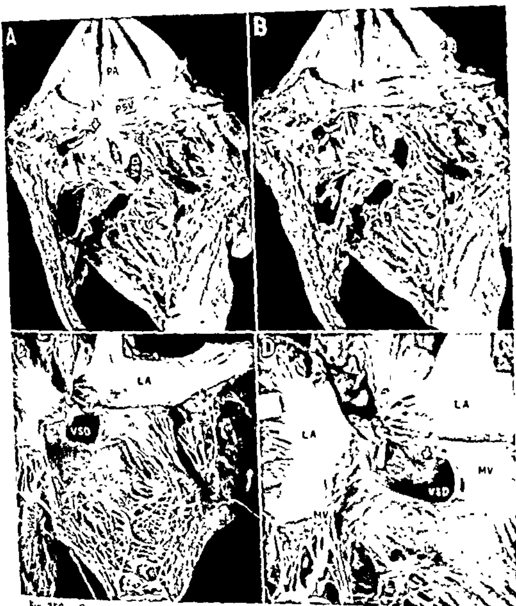


Fig 350 —Same case as in Figure 366 Woman aged 23 (L L 310201) [illegible]

[illegible] dorsally to this part of the crista, and opens directly into the right ventricle The *intracardial* segment of the aorta is short and the cusps of the aortic valve are attached at the level of the pulmonary valve The trabeculae of the *right ventricle* are greatly hypertrophied and their appearance is abnormal The [illegible] aorta is of somewhat less than the normal width A [illegible] the part of the posterior cusp of the mitral valve cl[illegible] placed right part of the infundibulum of the right ve[illegible] left ventricle are also hypertrophied CSV crista [illegible] left and right atria M[illegible] nary semilunar valves [illegible] tricular septal defect

Fig. 351a.—Large defect in membranous part of ventricular septum and 50 per cent over-riding of aorta. Girl, aged 9 months (Y P 520616) Injection of contrast medium into left ventricle through a catheter passed via the foramen ovale, pictures taken in 45° oblique projections. Septal defect lies between cusps of the aortic valve and muscular part of the ventricular septum (arrow in *B*); width is about [illegible] medium passes
into both right ventricle and a [illegible]
defect, partly because of the o[illegible]
lum of left ventricle, *LV* and *RV*, left and right ventricles, *VS*, ventricular septum



Fig. 351b.—Same case as in Figure 351a. In right ventricle (*A*), defect (arrow) is immediately above the medial cusp of the tricuspid valve and below the parietal band, which is thin, foreshortened and inserted far cranially. In left ventricle (*B*), defect (right arrow) is in the membranous part of the ventricular septum, immediately below the aortic valve. Its width is about ¾ of th
Con
of r
PB,

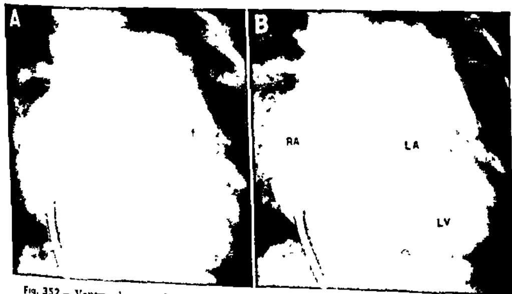


Fig. 352.—Ventricu
Catheter is inserted int
diluted by the large qu
left and right atria, *LV*

lated, owing to the increased blood flow in the pulmonary circulation. The large quantity of blood in the atrium causes great dilution of the contrast medium (Fig. 352). *Unfavorable conditions as far as the projections are concerned*, with partial overlapping of the relevant region of the ventricle by the contrast-filled atrium, may also mar the results.

When the contrast medium is injected intravenously, the recirculation to the right ventricle is the essential finding. Owing to dilution of the medium on passage through the pulmonary circulation, a study of the recirculation cannot be expected to provide satisfactory information regarding the anatomic conditions This fact has been fully substantiated by our experience.

In most cases, one is obliged to inject the contrast medium into the right ventricle If the septal defect is to be visualized by means of direct passage of the medium into the left ventricle, *the injection must be made so rapidly* that an instantaneous rise in pressure occurs in the ventricle and reverses the shunt With the present technique this method seems practicable only provided there is a definite rise in pressure in the right ventricle. With a systolic ventricular pressure of less than 50 mm Hg, we have only exceptionally succeeded in producing any direct passage of the contrast medium into the left ventricle. In the light of our experience, the injection should preferably be completed within 1 sec If possible, we have taken 12 exposures per second, otherwise six per second. This is because passage of the contrast medium through the defect could be induced only during diastole, and the rise in pressure caused by the injection is of short duration During systole, the pressure in the left ventricle usually *predominates*, blood which is not contrast-mixed then passes into the right ventricle and dilutes the contrast medium

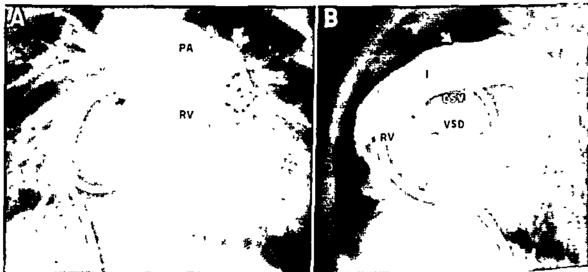


Fig. 353.—Defect in membranous part of ventricular septum Girl, aged 10 months (V U 530527). Contrast medium passes into the infundibulum of the left ventricle and outlines the lower surface of the aortic cusps (arrow in *A* and to the right of *CSV* in *B*). No apparent stenosis of the infundibulum of the right ventricle (arrow in *B* points to pulmonary valve) *CSV*, *crista supraventricularis*, *I*, infundibulum of right ventricle, *PA*, pulmonary artery, *RV*, right ventricle, *VSD*, ventricular septal defect

We consider that, in these investigations as well, frontal and lateral projections are preferable to oblique projections It is true *that the plane of the septum deviates from* the frontal plane in the thorax, and the image of the defect is distorted in both frontal and lateral views Theoretically, the defect is visualized without *distortion* in



Fig 354 —Small defect in membranous part of ventricular septum Girl, aged 8 (B-M.J 430527). Contrast medium spurts in a 3-4 mm wide jet into infundibulum (*I*) of left ventricle Defect lies between arrows RV, right ventricle

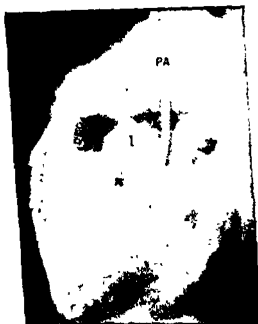


Fig 355 —Defect in membranous part of ventricular septum. Boy, aged 7 (L.B 481023) Defect 1 cm wide (between arrows) lies directly below crista supraventricularis (*CSV*) Good filling of infundibulum (I) of left ventricle and outlining of lower surface of aortic cusps PA, pulmonary artery

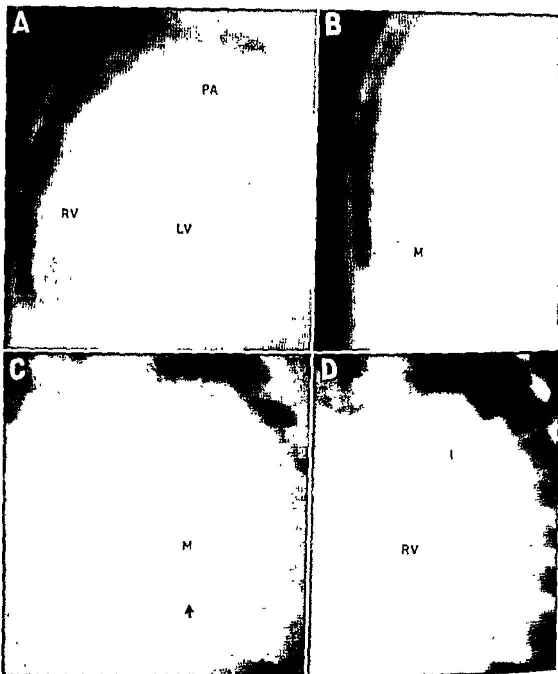


Fig. 356a.—Large defect in muscular part of ventricular septum and abnormal muscular ridge. Boy, aged 3 months (*L G 530425*) Great dilution of contrast medium, owing to large left [illegible] ng at the arrow in C and [illegible]), and its basal portion in [illegible] septal defect into the left ventricle. Right ventricle is large *and its* infundibulum wide, but the apical portion is small. *I*, infundibulum of right *ventricle*, *M*, muscular ridge, *PA*, pulmonary artery, *RV*, right ventricle.

one plane and axially in the other when oblique projections are used. But this implies that it can be visualized in one plane only, which makes it more difficult to determine the position of the defect. Moreover, the relation of the aortic root to the ventricular septal defect *is best judged* in the frontal and lateral projections. This also applies to evaluation of the position of

right ventricle passes into the subvalvula[r] region of the left ventricle, it become[s] dammed up below the aortic cusps durin[g] *diastole* (Figs. 351 and 355). During sy[s]tole, the contrast medium flows out of th[e] septal defect, and that part of it which wa[s] accumulated below the valve in the lef[t] *ventricle* is conveyed into the aorta

A septal defect in the *muscular part o[f]*

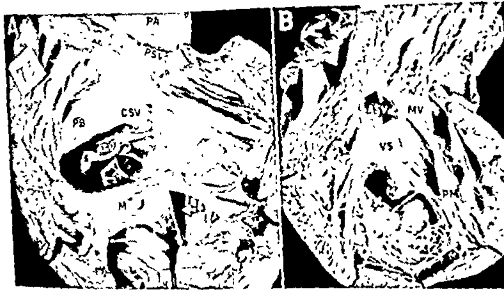


Fig. 356b.—Same case as in Figure 356a A, right, and B, left, ventricle Basal part of right ventricle is traversed by a thick muscular ridge (M), possibly consisting of maldevelo[ped] papillary muscles It runs from the basal part of the anterior wall, obliquely and backw[ard] [illegible]

[illegible] valves, TV, tricuspid valve, VS, ventricular septum.

the defect in relation to the crista supraventricularis

The findings on injection of the contrast medium *into the* right ventricle in the presence of a defect in the *membranous part of the septum* are illustrated in Figure 353 The septal defect can be identified in the right superior corner of the sinus region, below the crista supraventricularis, corresponding to the segment immediately below the aortic valve in the *left ventricle*.

When the contrast-mixed *blood from the*

the septum, lying more caudally to [the] crista supraventricularis, below and to [the] left of the membranous part of the sept[um] was visualized in two cases (Figs. 356a [and] 357) [illegible]

[illegible]

tricular septal defect with a mixed s[hunt] was made in 18 cases. Unquestion[ably] over-riding of the aorta could be dem[on]strated in only one case (Fig. 358). In others [illegible]

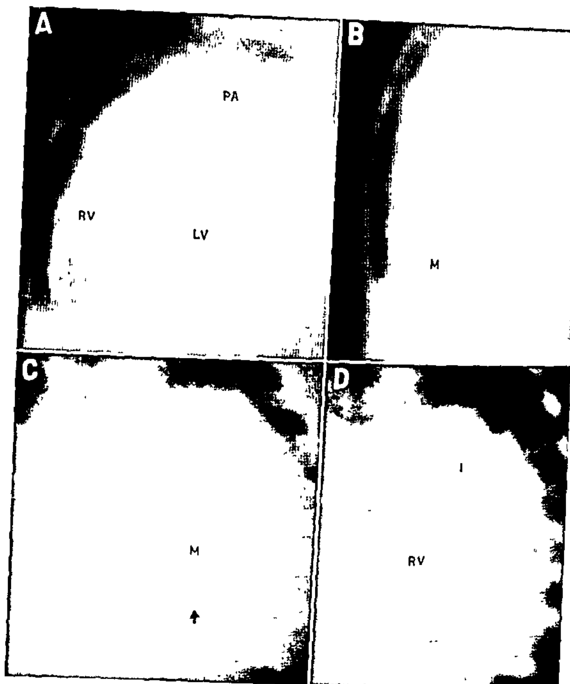
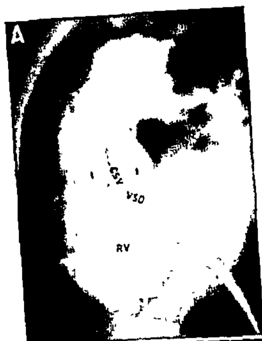
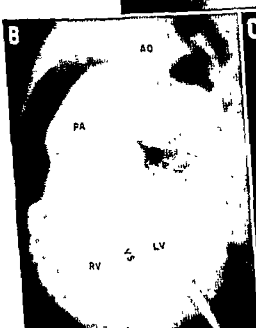


Fig. 356a.—Large defect in muscular part of ventricular septum and abnormal muscula
ridge *Boy, aged 3 months* (L.G 530425) Great dilution of contrast medium, owing to large le
to right shunt *There is a large filling defect in the right ventricle, starting at the arrow in C an*
extending upward The muscular ridge (M) is [illegible]

[illegible] 1, pulmonary artery, *RV*, right ventricle





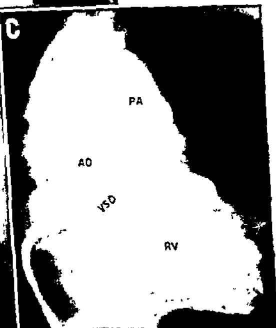


Fig 358.—Defect in membranous part of ventricular septum with equilibrated pressure in ventricles Girl, aged 2 (M A 520909) Wide defect, at the usual site in the right segment of the sinus region (*A* and *C*) Pulmonary artery distinctly dilated, aorta of normal width and about 50 per cent over-riding Greater part of the contrast medium passes into the pulmonary artery *AO*, aorta, CSV, crista supraventricularis, I, infundibulum of the ventricles, *LV* and *RV*, left and right ventricles, PA, pulmonary artery, VS, ventricular septum, *VSD*, ventricular septal defect.

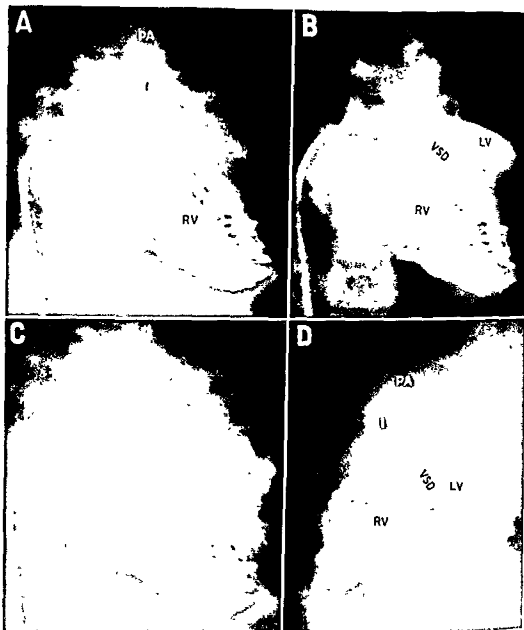


Fig. 357.—Large defect in muscular part of ventricular septum Girl, aged 3 (E W. 510707) Defect is caudal to the crista supraventricularis. A large quantity of contrast medium passes into left ventricle Trabeculae of the right ventricle are hypertrophied *I*, infundibulum of right ventricle, *LV* and *RV*, left and right ventricles, *PA*, pulmonary artery, *VSD*, ventricular septal defect

the aorta only through a wide ventricular septal defect situated either in the membranous or in the muscular part of the septum (Figs. 357 and 359–365). In most cases, the septal defect is not so sharply outlined by the contrast medium that its exact width can be determined Obviously, the width of the flow of the medium is no more than a minimum measurement.

latter event, the superior part of the ridge is displaced to the left and fused with the anterior wall of the ventricle close to the septum. Since the position of the inferior part of the ridge is normal, the septal defect lies in the superior part of the infundibulum, close to the pulmonary semilunar valves.

The following inferences may therefore



Late systole - - -
Early systole ———
Late diastole -·-·-



Late systole - - - - -
Early systole ———
Late diastole -·-·-·

Fig. 359b.— ... 359a *A*, fronta... left During sy... hypertrophied...

The conditions were unusual in the case illustrated in Figure 366. *Malformation of the dextrodorsal conus ridge* permitted filling of a large segment adjacent to the right part of the infundibulum and visualization of the aortic root, which was situated at an unusually high level and communicated directly with the outflow tract of the right ventricle. We have examined two other such cases.

The dextrodorsal conus ridge may be normal (Fig 367) or only partially malformed, as shown in Figure 370 In the

be drawn from the results of the angiocardiographic studies. In ventricular septal defect with a mixed shunt, the aortic root may be at the normal site. Moreover, frank overriding of the aorta, or displacement of the aortic root so that it lies side by side with the septum (as in Fig. 360), may occur in the presence of a left to right shunt only. As we have already stressed (p 339), these observations lend strong support to the view that, in subaortic septal defect, the direction of the shunt is determined mainly by the resistance in the pulmonary circulation

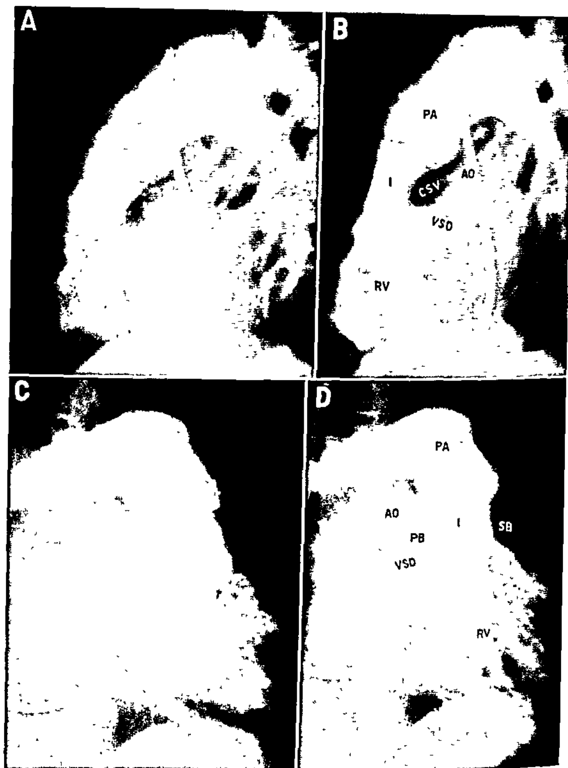


Fig. 359a.—Defect in membranous part of ventricular septum Girl, aged 9 (E S 440723). Infundibulum of right ventricle is slightly constricted, owing to forward displacement of right segment of the crista supraventricularis and the parietal band. The latter is foreshortened and inserted cranially. The constriction is not so severe as to be reflected in the pressure tracing The defect lies at the usual site. Contrast medium passes into the aorta, which is not over-riding, but lies side by side with the muscular part of the septum Pulmonary artery wide, aorta of normal width or somewhat narrower than normally, trabeculae hypertrophied *AO*, aorta, *CSV*, crista supraventricularis, *I*, infundibulum of right ventricle, *PA*, pulmonary artery, *PB*, parietal band, *RV*, right ventricle, *SB*, septal band, *VSD*, ventricular septal defect

A

B

C

PA

RV

LV

Fig 361 (*above left*).—Defect in membranous p[illegible]

520323). Defect [illegible]

cris [illegible]

pul [illegible]

Gir [illegible]

int [illegible]

aor [illegible]

[illegible], right ventricle

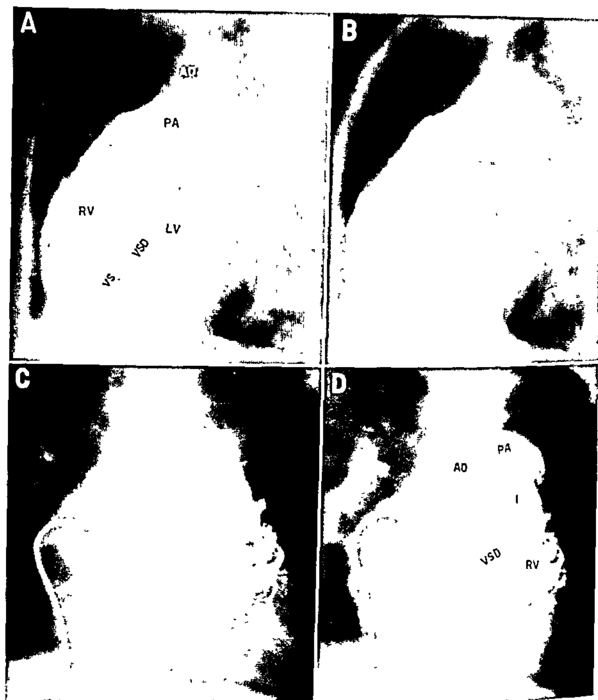


Fig. 360.—Defect in membranous part of ventricular septum with large left to right shunt and equilibrated pressure in ventricles Boy, aged 1 yr. (S K. 530513). Defect is at the usual *site* in the right segment of the sinus region *(D)* Because of rotation of the heart, relation of the aortic root to the septum is difficult to judge, it is probably not over-riding During systole *(C)*, ... *AO*, ... nc-









Fig 364 —Large defect in membranous part of ventricular septum Girl, aged 7 (I O 490916) The defect lies distinctly caudad to infundibulum (*I*) of both ventricles On catheterization, the catheter could be advanced into the aorta Mixed shunt Equalized pressure in ventricles Aorta narrow, AO, aorta, *LV*, left ventricle, *PA*, pulmonary artery, *RV*, right ventricle.



Fig. 363.—Large ventricular septal defect Boy, aged 12 (R K 430503) The defect involves the greater part of the septum Aorta (*AO*) narrow, not over-riding Aortic arch has abnormal course Mild coarctation of aorta Pulmonary artery (*PA*) wide *LV* and *RV*, left and right ventricles.

and to a lesser degree by the position of the aortic root

A defect in the membranous part of the ventricular septum combined with infundibular stenosis was studied by angiocardiography in eight cases. In several of them, our main object at the time of the investigation was not the visualization of the septal defect, but demonstration of the stenosis was observed in every case. In six cases, the infundibular stenosis was caused by the parietal band of the crista supraventricularis, which had an abnormal course. It compressed the ostium infundibuli from the right, from behind, and, to some extent, from in front as well In the two remaining cases, the stenosis was caused by both the parietal and the septal band A

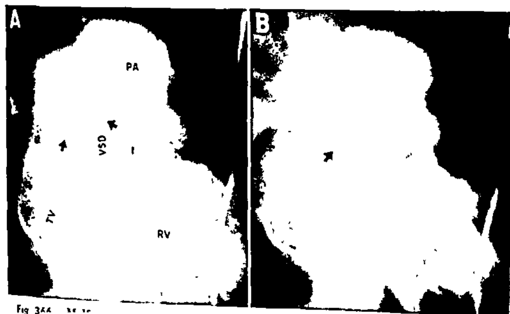


Fig 3[illegible] ure 350 which th outlined the pulm right ven

and elucidation of the anatomy of the pulmonary orifice Consequently, the contrast medium was not, in every case, injected with the rapidity necessary for identification of a septal defect

A septal defect at a typical site was demonstrated in three such cases (Figs 368 and 369) The aorta was not over-riding In several of the other cases there was distinct dilution of the contrast medium in the outflow tract of the right ventricle The filling defect was no more localized than in the other cases, consequently, it was not helpful in identification of the septal defect Recirculation to the right ventricle

case of this kind is illustrated in Figure 368 The infundibulum was large in all cases.

In two of the cases with a mixed shunt, the parietal band of the crista supraventricularis was slightly displaced to the left, causing inappreciable deformity of the ostium infundibuli, although this could not be detected on the pressure curve tracing. We did not classify this lesion as a stenosis.

A defect in the membranous part of the ventricular septum combined with valvular pulmonary stenosis was observed in three cases (Fig 371) In one, a patent ductus arteriosus was also present, its other as-



Fig. 365.—Large defect in membranous part of ventricular septum. Girl, aged 7 (S S 480417). *Defect* (between arrows) is about 1.5 cm in diameter Ostium infundibuli of right ventricle (*OI*) slightly constricted from both right and left side Width of aorta distinctly reduced *AO*, aorta, *I*, infundibulum, *LV*, left ventricle, *PA*, pulmonary artery, *RV*, right ventricle



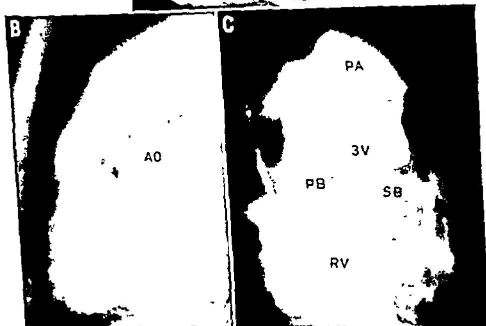
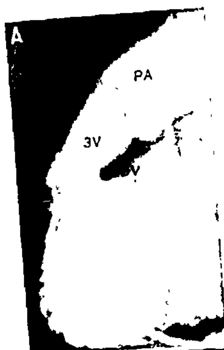


Fig 368 —Defect in the membranous part of the ventricular septum and infundibular stenosis Boy, aged 17 (T H 351219) Displacement of the septal and the parietal band, which bulge into the lumen and constrict the ostium infundibuli from the sides, from in front (arrow in A) and from behind (CSV) Large third ventricle, as wide as a normal infundibulum The defect is in the right superior part of the sinus, below the crista supraventricularis (arrow in *B*). Contrast medium is present in the infundibulum of the left ventricle (just above *PB* in *C*) and in the aorta (B), which is not over-riding *AO*, aorta, *CSV*, crista supraventricularis, *PA*, pulmonary artery, *PB*, parietal band *RV*, right ventricle, *SB*, septal band *3V*, third ventricle.



Fig. 367.—Small ventricular septal defect at site of infundibulum Boy, aged 4 (P 501110). A thin jet of contrast medium (arrow in *C*) spurts through the small defect into the infundibulum of the left ventricle Pressure in right ventricle only 30/0 mm Hg In *A* and distinct dilution of contrast medium (*x*) at site of septal defect





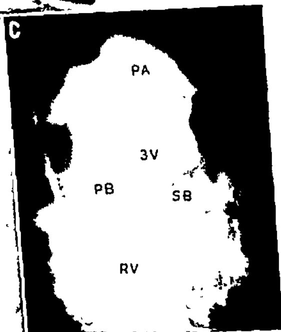


Fig 368 —Defect in the membranous part of the ventricular septum and infundibular stenosis Boy, aged 17 (T H 351219) Displacement of the septal and the parietal band, which bulge into the lumen and constrict the ostium infundibuli from the sides, from in front (arrow in [illegible] (*B*), which is not over-riding *AO*, aorta, *CSV*, crista supraventricularis, *PA*, pulmonary artery, PB, parietal band, *RV*, right ventricle, *SB*, septal band 3V, third ventricle

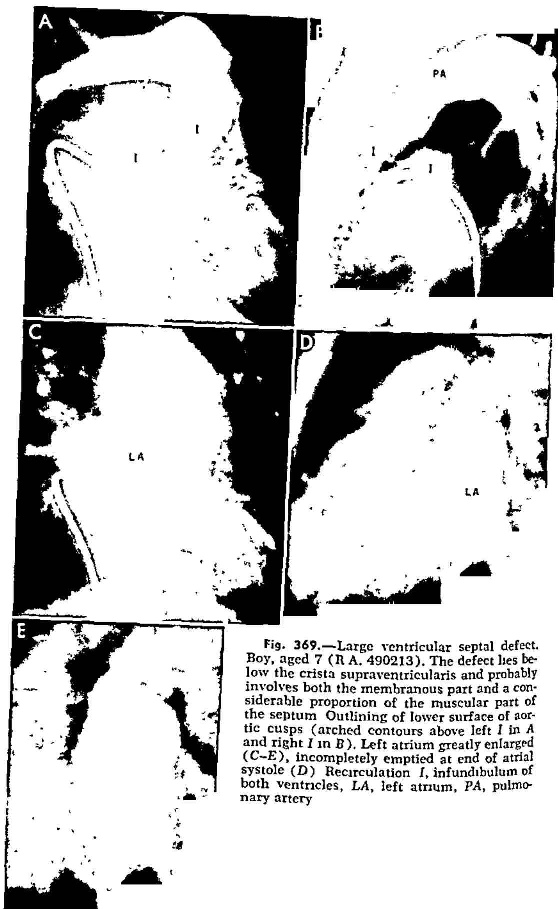


Fig. 369.—Large ventricular septal defect. Boy, aged 7 (R A. 490213). The defect lies below the crista supraventricularis and probably involves both the membranous part and a considerable proportion of the muscular part of the septum Outlining of lower surface of aortic cusps (arched contours above left *I* in *A* and right *I* in *B*). Left atrium greatly enlarged (*C*–*E*), incompletely emptied at end of atrial systole (*D*) Recirculation *I*, infundibulum of both ventricles, *LA*, left atrium, *PA*, pulmonary artery

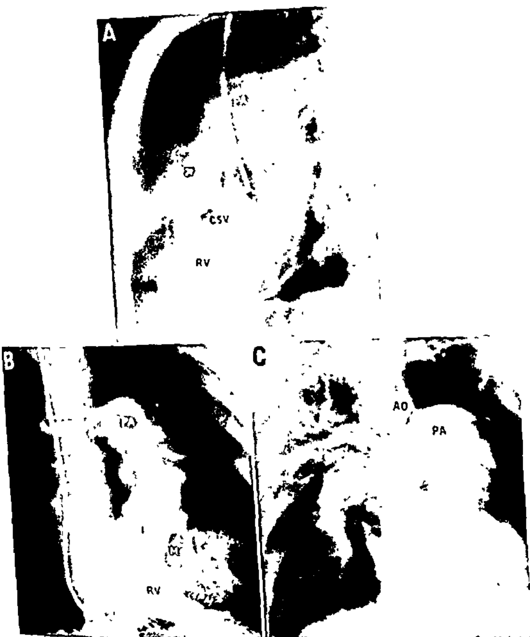


Fig 370a —Subvalvular ventricular septal defect with displacement of superior part of the dextrodorsal conus ridge Mixed shunt and equilibrated pressure in the ventricles Man, aged 37 (E M 170824) Defect, which is only partly filled with contrast medium, is immediately below the pulmonary valve (white arrow in *A* points to the valvular plane) and above the crista supra-

...channel is visible. Considerable dilatation of the pulmonary artery, extending to the main branches in the periphery, the vessels are narrow The aorta is narrow *AO*, aorta, *CSV*, crista supraventricularis, *I*, infundibulum, *M*, superior right segment of dextrodorsal conus ridge, *PA*, pulmonary artery, *PM*, papillary muscle, *RV*, right ventricle



Fig. 370b.—Same case as in Figure 370a Superior segment of dextrodorsal conus ridge is greatly displaced down and to left and fused with anterior wall of ventricle, close to septum (black-arrows in *A*) Between ridge and septum is a narrow channel (arrow, *B* and *D*; cut open in *D* and *E*) of appearance as in tetralogy of Fallot Large septal defect in right posterior part of infundibulum, close to the valve Caudal segment of dextrodorsal conus ridge is at the normal site but thin. Considerable hypertrophy of both the abnormal trabeculae and actual wall muscles in the right ventricle, which is large Anterior papillary muscle is displaced, originating in septum and fused with the septal band Pulmonary artery is grossly dilated, numerous atheromatous [illegible] semilunar valves. In [illegible] hangs down [illegible] narrower than normal *AO*, aorta, *ASV*, aortic semilu[illegible] [illegible]orsal conus ridge (right segment of crista suprav[illegible] [illegible] muscle, *PSV*, pulmonary semilunar valves, *SB*, sep[illegible] [illegible]ular septal defect (*continued*)

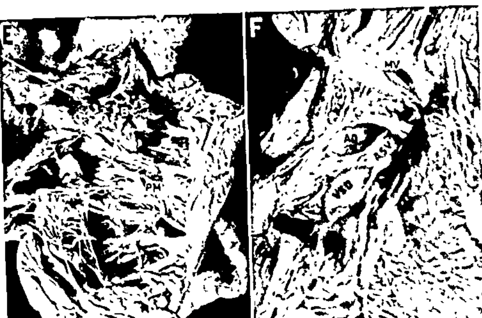


Fig 370b. (cont.)

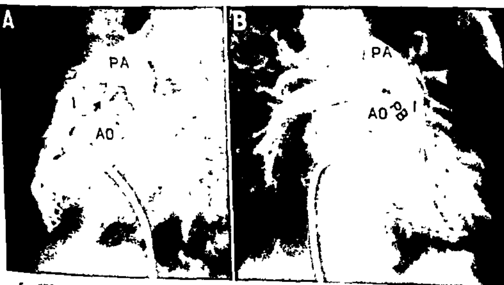


Fig 371.—Defect in membrano...
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(B)
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Fig. 370b.—Same case as in Figure 370a. Superior segment of dextrodorsal conus ridge is greatly displaced down and to left and fused with anterior wall of ventricle, close to septum (black-arrows in *A*). Between ridge and septum is a narrow channel (arrow, *B* and *D*, cut open in *D* and *E*) of appearance as in tetralogy of Fallot Large septal defect in right posterior part of infundibulum, close to the valve Caudal segment of dextrodorsal conus ridge is at the normal site but thin. Considerable hypertrophy of both the abnormal trabeculae and actual wall muscles in the right ventricle, which is large Anterior papillary muscle is displaced, originating in septum and fused with the septal band Pulmonary artery is grossly dilated, numerous atheromatous ... e normal semilunar valves. In ... hangs down

... (right segment of crista supra... ulmonary semilunar valves, *SB*, sep... t (*continued*)

Fig. 370b. (cont.)

Fig. 371.—Defect in membran…
and …
(B) I…
defec…
with … to the valvular stenosis No dilatation of the pulmonary artery *AO*, aorta, *I*, infundibulum, *PA*, pulmonary artery, *PB*, parietal band

pects are therefore discussed in Chapter 16, which is on this cardiac anomaly. Since the case is a good illustration of the angiocardiographic findings in ventricular septal defect with associated malformations, the angiocardiogram is shown here (Fig. 372). The septal defect is wide and lies at the typical site in the membranous part of the septum; the aortic root is normal. The valvular stenosis exhibits the characteristic appearance of an expanded membrane The patent ductus cannot, however, be identified on the angiocardiogram, it can only be surmised on the grounds of a diffuse dilution *of the contrast medium in the main* trunk of the pulmonary artery Judging by the findings on cardiac catheterization, no valvular stenosis was anticipated, since the systolic pressure was the same on both sides of the valve. After ligation of the ductus arteriosus, the stenosis could also be demonstrated hemodynamically on renewed catheterization.

An unusual finding consisting of a recess-like outward bulge of the infundibulum is illustrated in Figure 373.

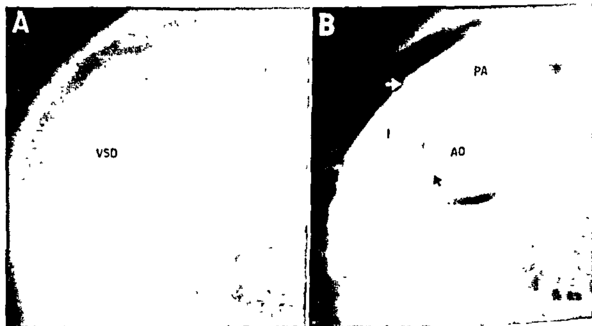


Fig. 372.—Defect in membranous part of ventricular septum, valvular pulmonary stenosis, and patent ductus arteriosus Girl, aged 4 months (B.K-S. 521028), see Figure 499 (p. 551). Defect is below the crista supraventricularis (black arrow in *B*), contrast medium passes into the infundibulum of the left ventricle and the aorta White arrow in *B* points to the valvular stenosis Pulmonary orifice is not visible Dilution of contrast medium in the pulmonary artery (*A*) affords evidence of an aortopulmonary communication *AO*, aorta, *I*, infundibulum, *PA*, pulmonary artery, *VSD*, ventricular septal defect.

VENTRICULAR SEPTAL DEFECT WITH TRICUSPID INCOMPETENCE

In ventricular septal defect combined with tricuspid incompetence, the findings on cardiac catheterization are similar to those in atrial septal defect (37). When the defect is in the membranous part of the septum, one of the cusps of the tricuspid valve may be fused to the septal defect and thereby prevent an interventricular shunt If the valve is fenestrated, a shunt to the right atrium occurs instead (310, 534). This is demonstrated by one autopsy case (Fig. 346) not belonging to our series and by one case in which both cardiac catheterization and angiocardiography were performed. In such cases, it is exceedingly difficult to make a clinical diagnosis.

Boy, aged 6 months (P T 520724) —The boy was mongoloid The heart disease was de-

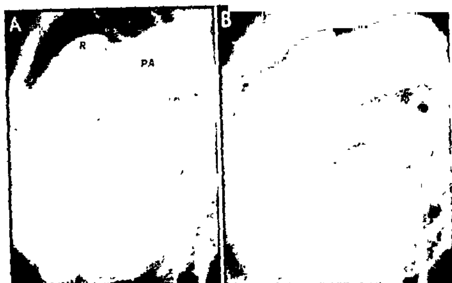


Fig 373 —Recess-like bulge in infundibulum of right ventricle Boy, aged 1 (O L. 550607) The recess (R), which shows marked contractility, overlaps the anterior aspect of the pulmonary artery

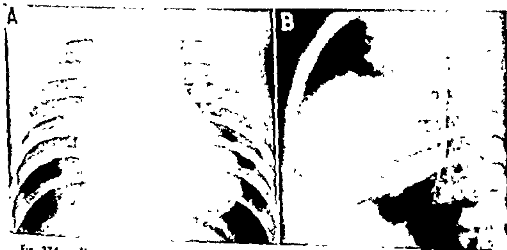


Fig 374a —Ventricular and atrial septal defect with tricuspid incompetence ... months (R.T. ...

tected at 2 months of age He had never exhibited cyanosis dyspnea or edema ...

... ventricular hypertrophy, incomplete right bundle-branch block, and tall P waves in V_1–V_4.

Roentgenologic examination (Fig 374a).— The heart was enlarged The right ventricle was dilated, and its anterior surface bulged as in hypertrophy The size of the left ventricle could not be estimated Both atria were enlarged The main trunk of the pulmonary ar-



Fig 374b (legend on facing page)

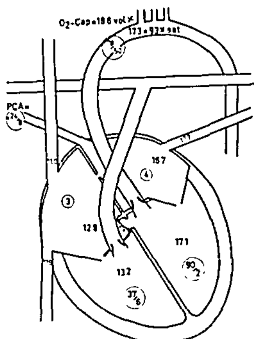


Fig. 375 —Ventricular septal defect with tricuspid incompetence. Schematic drawing demonstrating the hemodynamics Figures in circles denote systolic and diastolic pressure in mm Hg (in atria, the mean pressure), other figures denote oxygen content in volumes %

tery and its branches in and peripheral to the hilum were dilated, and there was increased vascularity of the lungs. The pulmonary artery exhibited lively pulsations The aorta was narrow.

Cardiac catheterization —The results are shown in Figure 375 The catheter passed from the right to the left atrium and was then advanced into the left ventricle and into the aorta It was not possible to pass the catheter from the right ventricle into the left ventricle or the aorta The pulmonary artery could not be catheterized Recording of the PCA pressure showed a characteristic, undamped arterial pulse curve with systolic pressure somewhat lower than that in the right ventricle, in which the pressure was only slightly raised Pulmonary stenosis could therefore be ruled out The pressure in the right atrium was normal, with no abnormal rise during ventricular systole Gas analysis showed the passage of oxygenated blood to the right atrium and of somewhat more to the right ventricle These findings were in entire agreement with those usually present in atrial septal defect. Moreover, there was presumably a small right to left interatrial shunt It is true that the mean pressure was higher in the left atrium than in the right, but during inspiration the fall was greater in the former, and a right to left shunt could then arise

←Fig 374b.—Same case as in Figure 374a Small atrial septal defect (left arrow in A and arrow in B) and large, oval defect in membranous part of the ventricular septum (C, D and arrow in E) Most of the ventricular septal defect is covered by the [illegible]

were found to be dilated, as were the pulmonary artery and its branches. Recirculation to the right ventricle occurred while contrast medium was still present in it and in the right atrium. These findings could be explained on the grounds of a ventricular septal defect, but were not proof of its existence.

On the second examination, in which the contrast medium was injected selectively through a catheter with the tip in the left atrium, the main part of the medium was emptied into the left ventricle. A small quantity leaked into the right atrium, but no communication between the atria could be clearly visualized. The enlarged left atrium overlapped the basal region of the ventricle, and the supposed ventricular septal defect could not be identified. The filling proceeded rapidly, and to a relatively greater degree in the right ventricle than in the right atrium We considered the results to warrant the assumption of an atrial septal defect, but it was regarded as uncertain whether or not a ventricular septal defect was also present.

Subsequent course.—The patient was sent home. At a check-up one year later, the heart was found to have undergone further enlargement. It was therefore decided to repair the atrial septal defect At operation, only a small atrial septal defect was found, which did not suffice to explain the clinical features. When the defect was closed, the right atrium dilated further The patient died in connection with the operation.

Autopsy (Fig 374b) disclosed a grossly enlarged heart with dilatation of both ventricles, but still greater dilatation of the right atrium The foramen ovale was not covered by the valve, but had an opening about 5 mm in diameter. The medial cusp of the tricuspid valve was fused to the ventricular septum and covered a defect in the membranous part of the septum The cusp was rudimentary, the chordae tendineae were lacking, and it was perforated with small holes which allowed passage of blood from the left ventricle to the right atrium. The other cusps were normal.

A shunt from left ventricle to right atrium may take place even if the cusps of the tricuspid valve are normal. This is because part of the membranous portion of the septum—the so-called atrioventricular portion—forms a dividing wall between the left ventricle and the floor of the right atrium. A defect in this portion of the septum results in direct communication between left ventricle and right atrium (216, 250). We have no such case in our series

DEFECT INVOLVING THE ENTIRE VENTRICULAR SEPTUM (SINGLE VENTRICLE)

As a hemodynamic conception, a single ventricle implies that both the aorta and the pulmonary artery take their origin from a common ventricle. Anatomically, there is not necessarily a single ventricle. The second ventricle may be present as a rudimentary cavity. This often applies in tricuspid or mitral atresia. In such cases, the right or the left ventricle consists of a small outlet chamber or of an entirely closed, nonfunctioning cavity. Single ventricle in the anatomic sense denotes a condition in which the two ventricles form a common chamber, owing to complete absence of the septum. Even if remnants of the septum are present but the defect is so large that the blood in the ventricles is entirely mixed, one is justified in speaking of a single ventricle.

In the majority of cases in which the ventricular septum is lacking, lesions of the atrioventricular valves are also present. Such cases in our series are reported in the relevant chapters, i.e., tricuspid stenosis (A A. 380728) and mitral atresia (E.L 520823). The patients usually present transposition of the great vessels as well (216).

Single ventricle with normal atrioventricular valves can be regarded as a special group among the ventricular septal defects Presumably, some remains of the septum exist, but the defect is so great that the blood from the two ventricles is completely mixed. We had two cases in our series The salient features were as follows.

Boy, aged 16 (O H 391109) —The patient also had slight valvular pulmonary stenosis, transposition of the great vessels, and absence of the inferior vena cava, the lower part of the body being drained by the hemiazygos vein. He had been cyanotic since the age of 2 months [illegible] as well as an accentuated secon[illegible] the pulmonary area Blood samples from the


A
C
D

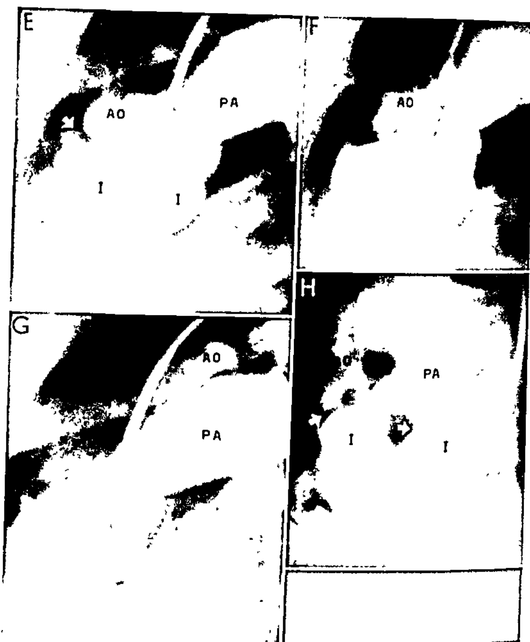


Fig. 376 (*cont*)



Fig 377.—Single ventricle Boy, aged 6 (C-G L 491103) A small septum is visible be-

right ventricle and from systemic arteries had the same oxygen saturation (83 per cent).

Roentgenologic examination.—This showed situs inversus. The heart was slightly enlarged The main trunk of the pulmonary artery was dilated, the ascending aorta ran on the right side, and the aortic arch produced a slight impression in the dorsal aspect of the trachea and esophagus. The descending aorta was visible on the left side The vascularity of both lungs was greatly increased

Angiocardiography —The contrast medium was injected into a chamber from which both the wide pulmonary artery and the narrower aorta were given off in transposed position (Fig. 376) Stenosis was present in the outflow tract of the aorta and pulmonary artery The inferior vena cava was lacking, and the catheter was introduced into the heart through the hemiazygos vein

Boy, aged 6 years (C-G L 491103) — Transposition of the great vessels was also present The child had only slight cyanosis, but had always been severely disabled, particularly by dyspnea. The second sound was accentuated over the pulmonary area, but no pathologic murmur was present. The oxygen saturation of blood samples from the right ventricle and aorta was the same (78 per cent). The *electrocardiogram* showed large amplitudes, but no ventricular hypertrophy.

Roentgenologic examination —This showed no enlargement of the heart or any characteristic change in shape. The left vascular outline was prominent and had an unusually lengthy course Absence of the typical impression of the aortic arch in the esophagus and trachea, as well as the course of the descending aorta *in the midline in front* of the spine, indicated that the aforementioned vascular outline might have represented the ascending aorta The main trunk of the pulmonary artery could not be identified The central and peripheral pulmonary vessels were greatly dilated.

Angiocardiography —The contrast medium was injected into the ventricle It is seen from Figure 377 that the aorta and pulmonary artery are given off from the same chamber, the aorta on the left, somewhat anterior to a greatly dilated pulmonary artery No remains of the septum could be identified in the ventricle, except between the two infundibula

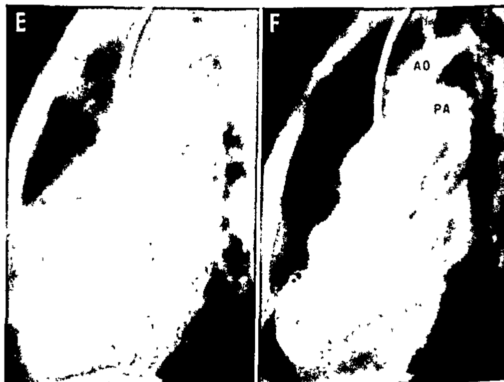


Fig. 377 (*cont*)

12 | Atrial Septal Defect

A PATENT foramen ovale is present in 20 to 25 per cent of all adults (51, 216). In such cases, the septa are fully developed and the valve covers the opening, but is imperfectly fused. In normal individuals, the pressure in the left [illegible]

ciation with certain pathologic conditions of the pulmonary circulation or of the right side of the heart, which cause a rise in pressure in the *right atrium*, that the foramen ovale may open and a right to left shunt develop.

By atrial septal defect is *meant* an opening between the atria which is not covered by the valve and which therefore permits blood flow in both directions. *If no other* abnormalities are present in the heart or the pulmonary circulation, a left to right interatrial shunt arises, with increased and ineffective blood flow through the right atrium and ventricle, the pulmonary circulation, and the *left atrium*. This part of the circulatory system becomes dilated, except for the left atrium, which is of normal size because it is drained into the right atrium through the defect, during both systole and diastole. In severe cases, there is diminution of the flow through the left ventricle and the systemic circulation and an increase in the arteriovenous oxygen difference.

When there is a complicating cardiac malformation, the picture may be entirely different. Extreme examples are atresia of the tricuspid or mitral valve, when only the presence of the defect is compatible with life. Consequently, it is essential to confine the clinical conception of atrial septal defect to entirely uncomplicated cases or those associated with only slight cardiac malformations which have little influence on the clinical features. The most common of them are mitral stenosis (Lutembacher's syndrome) and mild pulmonary stenosis. The classification of cases of combined pulmonary stenosis and atrial septal defect has been discussed (p. 138). We have classified cases with such mild pulmonary stenosis that a left to right shunt is present as atrial septal defects. Pulmonary hypertension may cause changes in the clinical picture, but since the raised pressure is so intimately associated with the shunt, these cases will be discussed with the uncomplicated ones.

ANATOMY

The anatomy of the two atria differs greatly (see p. 64) and partly explains the hemodynamics in atrial septal defect. The left atrium has a thick muscle wall, and only the *appendage* is trabeculated. The right atrium, in contrast, has an extensive trabecular *network*, and the wall between the trabeculae is exceedingly thin (Fig. 67, p. 67). Consequently, the right atrium more easily becomes dilated. In



Fig. 3[illegible] ...sorption of septum primum and possibly [illegible] *C*, underdevelopment of [illegible] 440828). Arrows point to the foramina venarum minimarum *D*, underdevelopment of septum secundum (J W. 450227). *E* and *F*, perforated valve with absorption at abnormal sites, underdevelopment of septum secundum and abnormally large foramen ovale (K J 1897156). Arrow points to Chiari's net. *G* and *H*, underdevelopment of septum secundum and absorption into septum primum at an abnormal site (K.J. 1895123) *I*, underdevelopment of septum secundum and over-resorption of septum primum (K J. 189525) *FoO*, fossa ovalis, *IVC* and *SVC*, inferior and superior venae cavae, *LA* and *RA*, left and right atria, *LAA* and *RAA*, left and right auricular appendages, *MV*, mitral valve, *TV*, tricuspid valve, *VCS*, valve of coronary sinus, *VVC*, valve of vena cava, *PV*, pulmonary vein (*continued*)

... of the ... the rest of the ... trium is, however, normal (51). The ventricle is dilated, as is the pulmo- artery. In uncomplicated atrial septal t, the hypertrophy of the right ventricle involves mainly the bands of the crista supraventricularis and the trabeculae With coincident pulmonary hypertension or pulmonary stenosis, there is also thickening of the ventricular wall

Both the size and the site of the atrial septal defect vary considerably (Figs 378–384), depending on the nature of the developmental anomalies of the septum primum and septum secundum (see Chap 1, p 19)



It is of great practical importance to establish the anatomy of the defect, since different surgical techniques are currently used for repair of different kinds of atrial septal defects. Classification of the defects should take these surgical considerations into account.

In conformity with other authors (390, 428) we have adopted the following classification

A. Common atrium.
B Foramen ovale defect.
C Sinus venosus defect.
D Ostium primum defect.

Several defects of different type may be present in the same individual.

A COMMON ATRIUM.—The septum is absent, with the possible exception of small

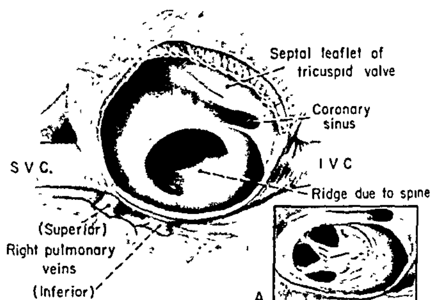


Fig. 379.—Figures 379 through 384 depict atrial septal defects from the surgeon's point view as he stands on the right side of the supine patient, with the patient's head to the left Th is a foramen ovale defect in which the valvula foraminis ovalis is absent. *A*, variation of t foramen ovale defect in which part of the valve remains as a fenestrated membrane (Fro Lewis *et al.*, Ann. Surg 142.401, 1955.)

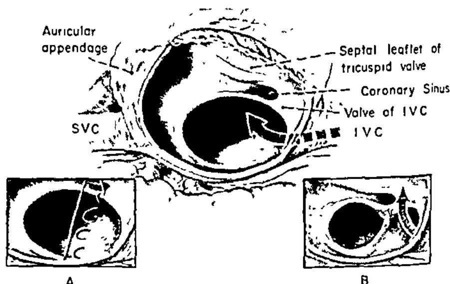


Fig. 380.—In this foramen ovale defect, the valvula foraminis ovale is absent, and th prominent valve of the inferior vena cava creates a false lower margin *A*, first repair stitch i place. *B*, first repair stitch tied (From Lewis *et al*, Ann Surg 142 401, 1955)

ridges along part of the margin of the common atrial chamber.

B FORAMEN OVALE DEFECT.—This type of defect arises either through a developmental arrest of the septum secundum or by over-resorption of the septum primum, which normally forms the valvula foraminis ovalis within the oval limits of the limbus (Fig 379) In the former case, the result is an opening too large to be closed by the septum primum, in the latter, the

C. SINUS VENOSUS DEFECT.—This has long been described as a special type (29, 273, 300, 332, 354, 664, 678), but it was not accorded any great interest until the introduction of surgical treatment of atrial septal defects (306, 308, 350, 430, 497a, 568, 686); see Figure 381. This defect has the following characteristics (1) it is situated above the fossa ovalis, (2) the septum has no upper or posterior margin, (3) the pulmonary vein from the upper lobe of the

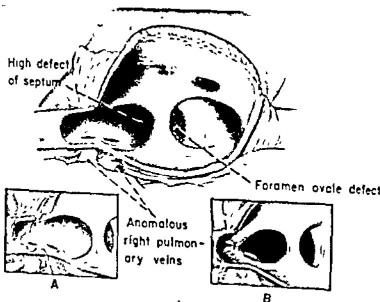


Fig 381.—...
pair stitch is p...
et al, Ann Su...

septum primum will no longer form a valve adequate to close the foramen ovale The latter condition seems to be the more common The defect may not only involve the fossa ovalis but extend toward the inferior vena cava, so that there is no lower margin of the defect above the orifice of this vessel (Fig 380) The blood can thereby be shunted more easily from the inferior vena cava to the left atrium (52, 366). On closure of the defect, there is a risk that the inferior vena cava will open into the left atrium instead of the right (366, 429) It is therefore important to be aware of this type of defect

right lung, and sometimes that from the middle and lower lobe as well, is connected with the superior vena cava at its opening into the right atrium. According to Ross (568) the defect may lie within the orifice of the superior vena cava, in its medial wall It sometimes extends as far as the fossa ovalis (Fig 382)

D OSTIUM PRIMUM DEFECT —This type of defect lies in the lower part of the atrial septum (Fig 383) The atrial septum has no margin above the ventricular septum. The defect is attributable to lack of fusion of the components of the atrial septum to the atrioventricular endocardial cushions

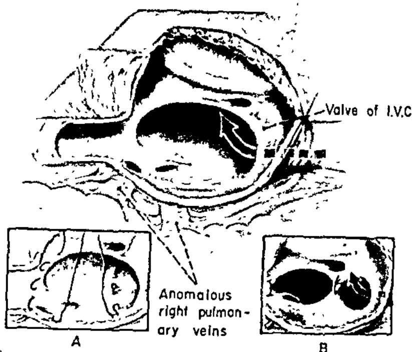


Fig. 382.—Large defect which has the characteristics of both a high defect and a foramen ovale defect. *A*, the upper stitch is the same as the first repair stitch of a high defect. The lower stitch is identical with the first repair stitch of the type of foramen ovale defect as shown in Figure 380. *B*, upper and lower stitches tied. (From Lewis *et al*, Ann Surg. 142 401, 1955)

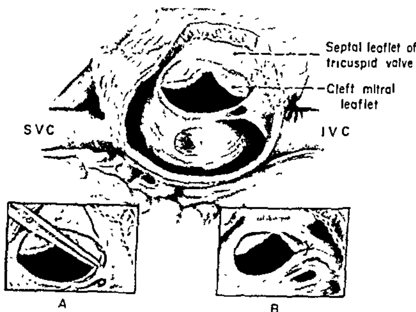


Fig. 383 —Persistent ostium primum with closed foramen ovale *A*, repair of this defect employing the lowermost part of the left atrial wall to avoid injuring the conduction system *B*, first stitch tied. (From Lewis *et al*, Ann Surg 142 401, 1955.)

and represents a persistence of the interatrial foramen primum (see pp. 19 and 22). This lack of fusion is dependent on a disturbance in development of the endocardial cushions

All transitional forms are found between this type of atrial septal defect and a common atrioventricular canal (Fig 384) with a defect involving the lower part of the atrial septum and the upper part of the ventricular septum and with a common view of the fact that there is an indefinite transition to a common atrioventricular canal, it might be desirable to assemble the defects in one group. Weidman *et al.* (691) and Wakai and Edwards (680) use the term persistent common atrioventricular canal and distinguish between a partial and a complete type. Campbell and Missen (138) use the term endocardial cushion defects and divide the defects into three types, as follows. Grade I: This defect "usually

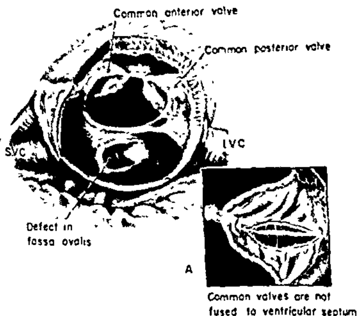


Fig 384.—Common atrioventricular canal with small foramen ovale defect A, detail of dorsal, common atrioventricular valve, which has been split to show how it is suspended above the ventricular septum by numerous small chordae (From Lewis *et al*, Ann Surg, 142 401, 1955)

atrioventricular valve. Even in cases with complete division of the atrioventricular canal and intact ventricular septum, malformations of the atrioventricular valves are usually present They are commonly in the form of a bifid anterior mitral cusp and sometimes in the form of a malformed septal tricuspid cusp. Occasional cases with normal valves have, however, been described (70), we have seen such a case, verified at autopsy, and another in which no valvular deformity was found at operation under direct vision in the open heart

From the embryologic standpoint, and in consists of a persistent ostium primum and a bifid anterior mitral cusp. Division of the A-V canal occurs, and the tricuspid valve is commonly normal, though occasionally it may be malformed instead of—or even as well as—the mitral valve" Grade II. "Here the dorsal and ventral cushions have just succeeded in fusing, as shown by the narrow bridge of fibrous valvular tissue attached to the upper surface of the ventricular septum, but the fusion has been insufficient to allow the formation of either the anterior leaflet of the mitral valve or the septal leaflet of the tricuspid valve, so that

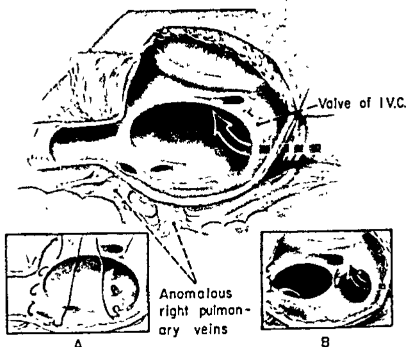


Fig. 382.—Large defect which has the characteristics of both a high defect and a foramen ovale defect. *A*, the upper stitch is the same as the first repair stitch of a high defect. The lower stitch is identical with the first repair stitch of the type of foramen ovale defect as shown in Figure 380. *B*, upper and lower stitches tied. (From Lewis *et al*, Ann. Surg. 142 401, 1955)

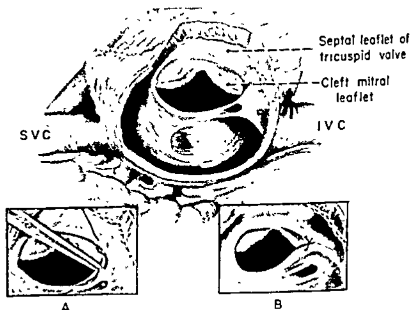


Fig. 383.—Persistent ostium primum with closed foramen ovale *A*, repair of this defect employing the lowermost part of the left atrial wall to avoid injuring the conduction system *B*, first stitch tied (From Lewis *et al*, Ann Surg 142 401, 1955)

mm Hg in the mildest case With a very large left to right shunt and a large flow through the pulmonary orifice, a pressure gradient of up to 20 mm Hg can be recorded even though no anatomic stenosis is present. This has been discussed in detail on page 120. In three cases the systolic pressure in the pulmonary artery was 50 mm Hg or above. In one case, interpreted earlier as atrial septal defect with pulmonary hypertension, a persistent ostium atrioventriculare commune could be diagnosed on angiocardiographic examination two years later

Complicating mitral stenosis increases the left to right shunt and aggravates the condition The mitral stenosis is probably of rheumatic origin Bedford *et al* (51) have stated that in 25 per cent of all cases of *atrial septal* defect there is associated mitral stenosis, and that this applies to women in particular According to Edwards (216), the diagnosis of Lutembacher's syndrome has been groundless *in many of the* cases reported, since the thickening of the cusps, which is so often observed in adults, has been misinterpreted as mitral stenosis If the origin is rheumatic, the incidence must vary considerably in relation to the geographic incidence of rheumatic *fever* In Sweden, this illness has been extremely uncommon during the past decade Our series includes only one case of Lutembacher's syndrome, verified at operation The patient was a 6-year-old girl (R J 461030) she had not had rheumatic fever

CLINICAL FEATURES

Taussig (650) has stated that patients with atrial septal defect often have a characteristic appearance Their build is frail, they are underdeveloped, and puberty is delayed The skin is *thin and* transparent In our experience, this picture is also seen in other conditions with an extremely large left to right shunt and decreased systemic blood flow (*ventricular* septal defect and patent ductus arteriosus) Figure 385 shows the physical development and age distribution of our patients It is true that most of them are normally developed, but there are many below the norm, and among the latter are patients with a large shunt or pulmonary hypertension.

The clinical picture varies with the size of the defect It ranges from severe symptoms as early as the first year of life (196, 677) to no symptoms even at a fairly advanced age (161) Death during the first year of life is, however, considerably more unusual than in ventricular septal defect (139, 319). The increased burden on the right ventricle gradually leads to congestive failure, but not, as a rule, until the third to fifth decade (51, 52, 139, 561). As right ventricular failure develops, *the direction* of the shunt changes and cyanosis appears (187). In contrast to the findings in the majority of congenital heart diseases, bacterial endocarditis is rare in cases of atrial septal defect (216)

Forty-one of our patients were entirely asymptomatic. In the others, the most usual symptom was increased fatigability, the next in order being dyspnea Slight peripheral *cyanosis* on exertion was found in eight Cyanosis is not necessarily central, but may be explained on the grounds of the reduction in systemic flow and the resulting increase in the arteriovenous oxygen difference Particularly during the first year of life, respiratory infections are strikingly frequent

In one of our cases, heart failure appeared at 2½ years of age Cardiac catheterization had been performed five months *earlier* An extremely large left to right shunt was found and fairly normal mean *pressure in* the pulmonary artery, but a large pulse amplitude, the systolic pressure was 35 mm Hg and the diastolic pressure 6 mm Physical signs of mitral regurgitation were present, and operation disclosed an ostium primum defect with cleft mitral valve.

Physical Working Capacity

The physical working capacity in adults with atrial septal defects has been *studied* at the department of Clinical Physiology,

both valves appear to have been incompetent: no interventricular communication remains, but there is a large persistent ostium primum." Grade III: "If fusion of the dorsal and ventral cushions fails entirely, the common A-V canal persists, and with it the ostium primum and a high ventricular septal defect, the three deficiencies being continuous"

From the clinical viewpoint, the main consideration is to distinguish between defects which only join the two atria and those which also have an interventricular communication. We have classified the latter type as persistent ostium atrioventriculare commune and have accounted for such defects in Chapter 13. The former type has been assigned to the group of atrial septal defects under the heading of ostium primum defects. If the atrioventricular valves are normal, or so inappreciably malformed that no regurgitation of any functional consequence occurs, these defects produce the same clinical features as other types of atrial septal defect.

If significant regurgitation takes place through the atrioventricular valves, the clinical picture is no longer the same. A distinction should therefore be made between ostium primum defects with mitral or tricuspid regurgitation and those without regurgitation.

It is possible, mainly with the help of angiocardiography and injection of contrast medium into the left atrium, to diagnose the various types of atrial septal defect with a fairly high degree of certainty. In our first series of cases, angiocardiographic examination was only exceptionally performed, consequently, the anatomy of the defect was not established in a large proportion of them Re-examination has been made in several of these earlier cases, permitting their proper classification. In two cases, which were earlier interpreted as being instances of partial anomalous drainage of the pulmonary veins, complementary examination revealed a sinus venosus defect, which could be verified at operation

Our series includes 77 cases of atrial septal defect, all studied by cardiac catheterization. In addition, 17 adults were examined roentgenologically (the remaining investigations, including cardiac catheterization, were performed at other clinics).

In 53 of these 77 cases, the defect could be classified according to the aforegoing criteria on the basis of the angiocardiographic features (see p. 450) and findings at operation (33 cases) or autopsy (4 cases). The distribution in the respective groups was as follows:

		No of Cases
A Common atrium		2
B Foramen ovale defect		33
a) In the region of the fossa ovalis.	31	
b) In the region of the orifice of the inferior vena cava	2	
C. Sinus venosus defect		6
D Ostium primum defect		12
a) Without regurgitation in the A-V valves	8	
b) With regurgitation in the A-V valves	2	
c) Uncertain regurgitation	2	

In two of the 12 cases with an ostium primum defect, the atrioventricular valves were normal, as shown by autopsy in one and operation (on the open heart) in the other. A cleft mitral valve without regurgitation was observed in one case at operation (on the open heart) and in another at autopsy. In four cases in which operation was performed with the closed technique, no regurgitation was palpable, but this does not necessarily rule out the presence of slight malformation of the valves. Only in two patients not yet operated on were there physical signs of regurgitation.

A large preponderance of women has been found in other series (562). In our material, 43 patients were girls and 34 boys

Complicating cardiac anomalies were present in nine cases, i e., mitral stenosis (one case), mild coarctation of the aorta (one case), valvular pulmonary stenosis (six cases), and infundibular pulmonary stenosis (one case). Pulmonary stenosis could be demonstrated by angiocardiography, despite a pressure gradient of only 19

Karolinska Sjukhuset (366). In patients under 25 years of age, the working capacity was usually normal despite a large left to right shunt Patients over 25 years old, on the contrary, had a low working capacity. Ten children aged 6 to 17 years had exercise tolerance tests, the working capacity was decreased in only one of them, although several had an extremely large shunt Four patients with normal working capacity in the test were stated in the history to be markedly disabled by their heart disease Subjective complaints often make their appearance, in fact, when the patients are informed of the presence of heart disease.

Physical Signs

Atrial septal defect as an isolated malformation with a left to right shunt is characterized by signs of right ventricular dilatation, a *systolic murmur* over the pulmonary area, a split second sound and, if [illegible]

[illegible]

An analysis of our 77 cases gave *the following* results

Palpation of the precordium.—*Precordial* heave was found *in most cases* In all four cases with pul*monary hypertension* it was conspicuous In the others it was *only faintly* discernible or at most moderate Roesler (562) has pointed out that the right ventricle may be displaced far *to the* left by a greatly enlarged right atrium The right ventricular lift toward the thoracic wall *is* then palpated so far to the left that it may be difficult to distinguish from an increase in the breadth of the apex beat in left ventricular *hypertrophy* The pulsations in the dilated *pulmonary* artery can sometimes be palpated (713) In severe cases, the peripheral artery pulse is small Increased *pressure* in the right atrium may result in dilatation of the veins

Reinbold and Nadas (554) have stated that the phlebogram is characteristic, with a marked *v* wave of approximately the same size as the *a* wave.

Sounds and murmurs.—In our cases, the first heart sound was normal. An early systolic pulmonary sound (417) could be demonstrated with certainty in only three cases, and in two of them there was associated pulmonary hypertension (see Fig. 389). In an additional three cases, one of them with pulmonary hypertension, the occurrence of this sound *phenomenon was* regarded as somewhat doubtful Thus, the sound phenomenon in question does not seem to be a regular feature of those cases of dilatation of the pulmonary artery that are not combined with pulmonary hypertension (416, 417), except in mild valvular pulmonary stenosis. The question of why this systolic click is so often found in cases of idiopathic dilatation of the pulmonary artery with normal pressure conditions does not appear to have been finally answered

Wide splitting of the second sound was a *constant finding* in all our cases with a *moderate or large* shunt without pulmonary hypertension. In the three cases with pulmonary hypertension, it was not possible to distinguish between the two components of the second sound (Fig 389). In atrial [illegible]

[illegible] *cardiographically, the interval* between the aortic and pulmonary components varied *from 0.04 to 0.08* sec. The intensity of both *components* was about the same

The *typical systolic murmur* is another important sign. In all uncomplicated cases, *it has a distinct* maximum over the pulmonary *area* It ranges from faint to moderately *loud* and is, as a rule, *considerably* fainter than in ventricular septal defect and pulmonary *stenosis (417)*. It is therefore seldom accompanied by a *thrill*. It is early systolic in time.

Phonocardiographic studies have shown that the murmur exhibits great similarities to that *which can* be recorded in healthy children, *in* whom blood flow is increased during *exercise*. It is true that, during exercise, this takes place *principally* by means of an increase in the pulse rate, without any

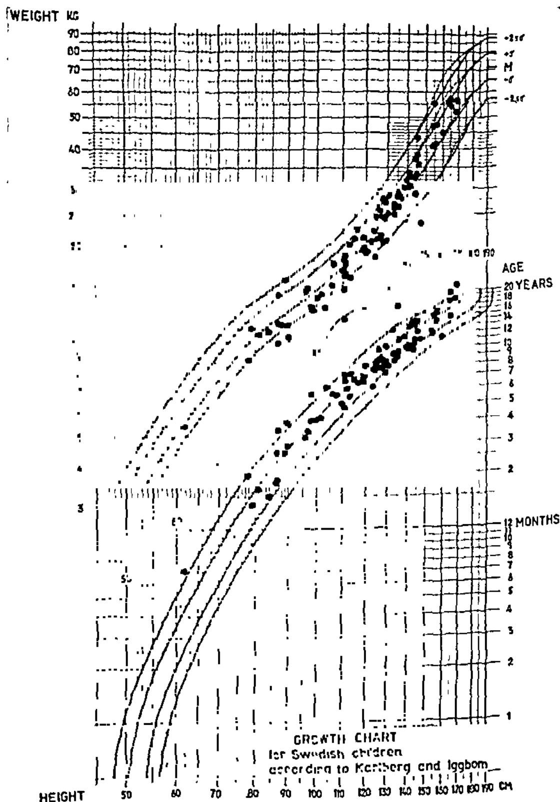


Fig. 385.—Physical development in 77 cases of atrial septal defect Height and weight majority of patients are within normal limits, several are below norm, among them, patie with large shunt or pulmonary hypertension (Relevant growth chart was kindly placed at disposal by Drs P Karlberg and S Iggbom [374, 375])

Karolinska Sjukhuset (366). In patients

the contrary, had a low working capacity. Ten children aged 6 to 17 years had exercise tolerance tests, the working capacity was decreased in only one of them, although several had an extremely large shunt Four patients with normal working capacity in the test were stated in the history to be markedly disabled by their heart disease Subjective complaints often make their appearance, in fact, when the patients are informed of the presence of heart disease

Physical Signs

Atrial septal defect as an isolated malformation with a left to right shunt is characterized by signs of right ventricular dilatation, a systolic murmur over the pulmonary area, a split second sound and, if the shunt is large, an apical diastolic murmur as well (38, 51, 75, 107, 202, 415, 416, 481, 554, 650, 713).

An analysis of our 77 cases gave the following results

Palpation of the precordium —Precordial heave was found in most cases In all four cases with pulmonary hypertension it was conspicuous In the others it was only faintly discernible or at most moderate Roesler (562) has pointed out that the right ventricle may be displaced far to the left by a greatly enlarged right atrium The right ventricular lift toward the thoracic wall is then palpated so far to the left that it may be difficult to distinguish from an increase in the breadth of the apex beat in left ventricular hypertrophy. The pulsations in the dilated pulmonary artery can sometimes be palpated (713) In severe cases, the peripheral artery pulse is small Increased pressure in the right atrium may result in dilatation of the veins

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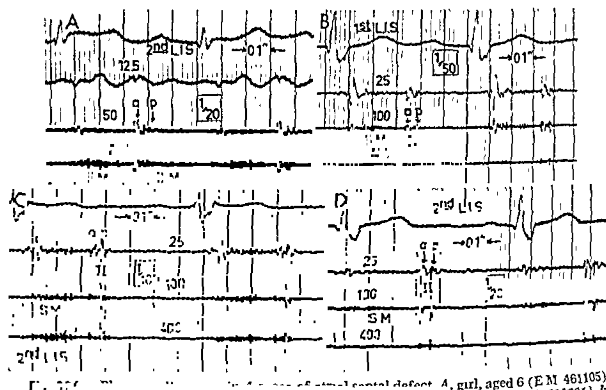
Wide splitting of the second sound was a constant finding in all our cases with a moderate or large shunt without pulmonary hypertension. In the three cases with pulmonary hypertension, it was not possible to distinguish between the two components of the second sound (Fig 389). In atrial septal defect, splitting of the second sound seems to be dependent on the low resistance in the pulmonary circulation (365) Phonocardiographically, the interval between the aortic and pulmonary components varied from 0 01 to 0 08 sec The intensity of both components was about the same

The *typical systolic murmur* is another important sign In all uncomplicated cases, it has a distinct maximum over the pulmonary area It ranges from faint to moderately loud and is, as a rule, considerably fainter than in ventricular septal defect and pulmonary stenosis (417). It is therefore seldom accompanied by a thrill It is early systolic in time

Phonocardiographic studies have shown that the murmur exhibits great similarities to that which can be recorded in healthy children, in whom blood flow is increased during exercise It is true that, during exercise, this takes place principally by means of an increase in the pulse rate, without any

notable increase in the stroke volume. It nevertheless implies a more rapid flow through the pulmonary orifice, since ventricular systole is also shortened. Figures 386 and 388 show the phonocardiograms in five cases of atrial septal defect. They should be compared with the recordings in Figure 387, which are taken from Mannheimer and Paulin's investigations (467) on the physiologic murmur at rest, after exercise, with a rise in the pulse rate, after inhalation of amyl nitrite, and on slowing of the pulse rate after administration of dihydroergotamine.

(We are able, partly on the basis of this experience, to share the view of Taussig (650) and of Blount *et al* (75) that the systolic murmur in atrial septal defect is not caused by the blood flow through the defect, but by "the increased blood flow through the pulmonary artery" (75). It has also been stated that the murmur is due to a "functional" or "relative" stenosis (51, 75)



F[illegible] of atrial septal defect. *A*, girl, aged 6 (E M 461105). [illegible] l, aged 10 (G S 411001). *In* [illegible] d splitting of the 2nd sound [illegible] denote degree of amplification, other figures denote standard frequencies of the filters *II a* and *b*, aortic and pulmonary components of 2nd sound, *DM*, diastolic murmur, *L I S*, left interspace.

The ejection time of the right ventricle is not prolonged in atrial septal defect, even though the stroke volume may be more than three times the normal one (365). This implies that the velocity of flow through the pulmonary orifice is so great that an intense murmur may arise. It is nevertheless difficult to draw any definite conclusions regarding the volume of the flow from the intensity of the murmur.) This is because a considerable role is played by the distance between pulmonary artery and thoracic wall (cf. Fig. 134, p. 148). It can be inferred from Figure 390 that the murmur may be louder in a healthy subject than in a patient with atrial septal defect. Particularly in an adult, the murmur may be faint despite a large shunt. A widely split second sound is a more constant sign. When there is coincident pulmonary stenosis, valvular or infundibular, the murmur has the same appearance as in these malformations (see p. 146).

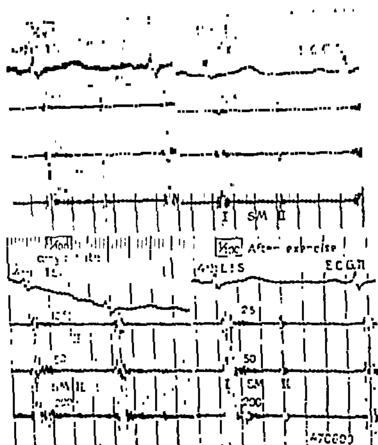
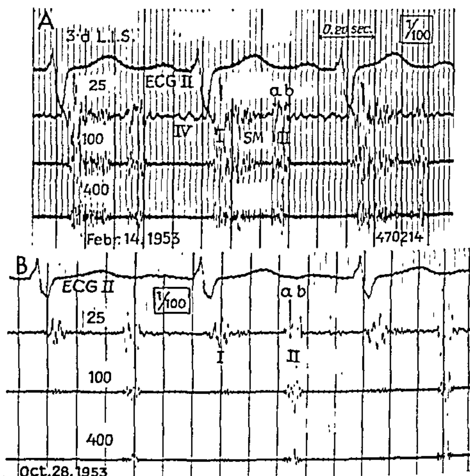


Fig 387.—Phonocardiograms of a healthy child. Boy, aged 6. Note the physiologic midsys-
ic murmur, it is best seen in the 200 frequency range at rest and after dihydroergotamine
HE). After both exercise and amyl nitrite, the systolic murmur becomes louder and protosys-
ic in time Note also the decreased amplitude of the 2nd sound, due to hypotension, after in-
lation of amyl nitrite Boxed figures denote degree of amplification, other figures denote
ndard frequencies of the filters II, 2nd sound, L I S, left interspace, *SM*, systolic murmur.
igs 387 and 388, from Mannheimer, E, in Levine, S Z [ed]. *Advances in Pediatrics* [Chi-
go Year Book Publishers, Inc, 1955], Vol VII)

A pansystolic murmur over the apex and in the left axilla, caused by mitral regurgitation, was heard in only two of our 12 cases classified as ostium primum defects.

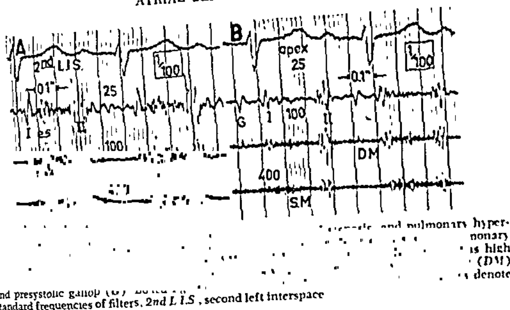
During the first month of life, a murmur is seldom heard (139, 569). Since during fetal life the flow passes in the normal way from right to left atrium, the right ventricle is of normal size at birth Some time elapses before it becomes dilated and the stroke volume increases sufficiently to produce a systolic murmur.

A diastolic murmur was heard in five cases over the pulmonary area (Fig 386, A) and was interpreted as a sign of pulmonary incompetence (Graham Steell murmur) In 32 cases there was a short, low frequency mid-diastolic apical murmur, probably caused by the rapid flow through the tricuspid orifice in the beginning of the filling phase of the right ventricle. Auscultation and phonocardiography do not seem to afford the possibility of distinguishing between cases with and without mitral stenosis (75, 503). Figure 389 is a phonocardiogram of a case of Lutembacher's syndrome (verified at operation) and pulmonary hypertension. The loud systolic murmur, the accentuated but not definitely split second sound, the apical diastolic murmur, and the auricular sound gallop are the salient features.



... A preopera-

filters II a and b, aortic and pulmonary components of 2nd sound, L I S, left inters... systolic murmur.



... stenosis and pulmonary hyper- ... nonary ... is high ... (DM) ... denote and presystolic gallop (G) ... standard frequencies of filters, 2nd L I.S, second left interspace

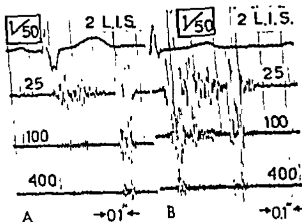


Fig 390.—Phonocardiograms A, in atrial septal defect Boy, aged 6 (C G 510526) The ratio of pulmonary to systemic flow was 2 1 B, in a healthy boy, aged 10 (K A 450408) The physiologic systolic murmur may sometimes be louder than the systolic murmur in atrial septal defect Boxed figures denote degree of amplification, other figures denote standard frequencies of the filters, 2 L I S, second left interspace

To sum up it may be stated that physical examination and phonocardiography usually lead to a correct diagnosis in uncomplicated atrial septal defect The characteristic findings are a fairly faint protosystolic murmur over the second left interspace, a split but not accentuated second sound, and frequently a diastolic murmur over the apex In addition, there is sometimes a parasternal lift displaced to the left and, in severe cases, a small peripheral artery pulse.

When the malformation is complicated by pulmonary stenosis, the murmur is louder If pulmonary hypertension supervenes, the second sound is accentuated and not definitely split In these cases, an early systolic pulmonary sound is usually heard and recorded

A pansystolic murmur over the apex and in the left axilla, caused by mitral regurgitation, was heard in only two of our 12 cases classified as ostium primum defects.

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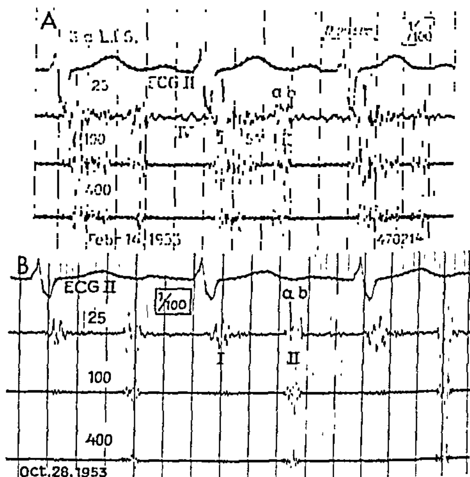


Fig 388.—Phonocardiograms in atrial septal defect Boy, aged 6 (S P. 470214). *A*, preopera tive, and *B*, postoperative examination. Note the fairly loud systolic murmur and split 2nd sound After operation, the murmur has disappeared and splitting of the 2nd sound is far less marked Boxed figures denote degree of amplification, other figures denote standard frequencies of th filters *II a* and *b*, aortic and pulmonary components of 2nd sound, *L I S*, left interspace, *SM* systolic murmur

from right to left atrium, the right ventricle is of normal size at birth. Some time elapses before it becomes dilated and the stroke volume increases sufficiently to produce a systolic murmur

A diastolic murmur was heard in five cases over the pulmonary area (Fig 386, *A*) and was interpreted as a sign of pulmonary incompetence (Graham Steell murmur)

between cases with and without mitral ste nosis (75, 503). Figure 389 is a phono cardiogram of a case of Lutembacher's syndrome (verified at operation) and pul monary hypertension The loud systolic murmur, the accentuated but not definitely split second sound, the apical diastolic murmur, and the auricular sound gallop are the salient features.

us defect with partial anomalous us return and no other malformations. diagnosis was confirmed at operation other patients had an ostium primum ct, one with mitral regurgitation and without. On the other hand, left axis ation was lacking in five patients with ostium primum defect, in one with sical signs of mitral regurgitation The gnosis could be verified at operation in e of these five cases.

Thus it is not possible by electrocardiog-

and Bedford, Papp, and Parkinson (51). Healey *et al.* (322) analyzed the correlation between the roentgenologic features and the changes in the hemodynamics in 12 cases of this cardiac anomaly.

The roentgenologic appearance is characterized by:

[illegible]

tricle.

A narrow aorta

Absence of dilatation of the left atrium.

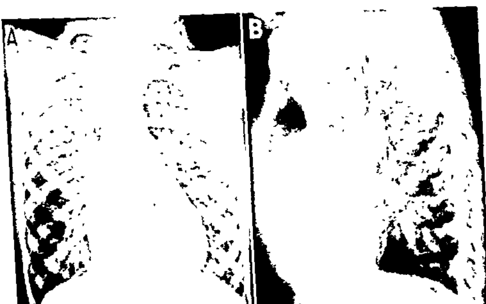


Fig 392 —Atrial septal defect Boy, aged 6 (S P 470214), see Figure 419 Considerable enlargement o[illegible] wall, norma[illegible] of pulmona[illegible] ment of left [illegible]

raphy to distinguish with certainty between an ostium primum defect and other types of atrial septal defect or between cases with and without mitral regurgitation However, if left axis deviation is present, the existence of an ostium primum defect is highly probable

ROENTGENOLOGIC EXAMINATION

Accounts of the main roentgenologic findings in atrial septal defect have been given by Assmann (25), Roesler (562, 563)

A roentgenologic examination was made in 91 77 children and adolescents and 17 adults (see p 416).

Dilatation of the main trunk of the pulmonary artery and of its central and peripheral branches (Fig 392) was the most constant feature and was present in every case The pulsations in the main trunk and its central branches were often large, as an indication of raised pulse pressure Aneurysmal dilatation of the pulmonary artery, which has been observed in adults, was not seen in any of the children. When there was

Electrocardiography

The electrocardiographic changes in atrial septal defect are confined to the right side of the heart. The left to right shunt places an increased burden on the right atrium and ventricle, which become dilated (diastolic overloading of right ventricle [126]) and, to some extent, hypertrophied. When there is associated pulmonary stenosis or pulmonary hypertension, the hypertrophy becomes more conspicuous whereas the dilatation is inappreciable since, with increasing right ventricular hypertrophy, between the degree of electrocardiographic changes and the pressure in the right ventricle, in conformity with the findings of other workers (604).

In some series, prolongation of the PR interval was recorded in up to 25 per cent of the cases (502, 616), but in our series it was recorded in only eight cases One patient had a congenital complete A-V block Tall, peaked P waves in V_1, often observed in severe pulmonary stenosis as a result of right atrial hypertrophy, are not as common in atrial septal defect when there is chiefly dilatation of the atrium. Slight P wave

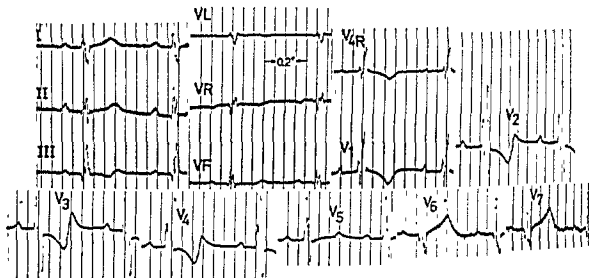


Fig. 391.—Electrocardiogram in atrial septal defect Boy, aged 14 (E B 400219). Incomplete right bundle-branch block, normal pressure in pulmonary artery, large left to right interatrial shunt

the left to right shunt becomes smaller (see p. 442). Dilatation of the right ventricle results in a prolonged activation time Incomplete right bundle-branch block, sometimes combined with right ventricular hypertrophy, is the most characteristic electrocardiographic finding in atrial septal defect (38, 673)

In our series only four patients had a normal electrocardiogram. An incomplete right bundle-branch block (Fig 391) was present in 58 cases, and a complete right bundle-branch block, in three Marked right ventricular hypertrophy was present in 13 cases and slight right ventricular hypertrophy in 31 No definite correlation existed anomalies were, however, seen in 15 cases Auricular flutter was present in two cases and was regularized by digitalis.

Left axis deviation in combination with incomplete right bundle-branch block, and sometimes signs of left ventricular hypertrophy, has been described as characteristic of ostium primum defects (69, 564, 656), even if these features are not constant (52) According to some authors, they are typical only of cases combined with mitral regurgitation (81, 502). In the frontal plane, the QRS loop rotates counterclockwise.

Left axis deviation was present in eight of our patients One of them had a sinus

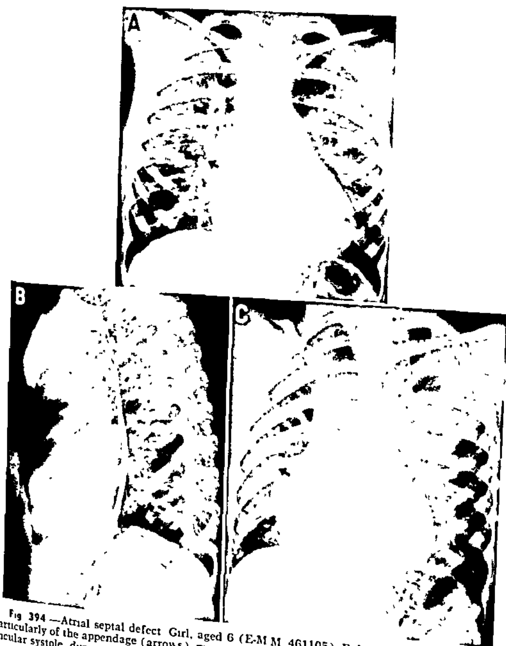


Fig 394 —Atrial septal defect Girl, aged 6 (E-M M 461105) Enlargement of right atrium, particularly of the appendage (arrows), most conspicuous in the left *oblique* projection *A*, ventricular systole, during which the atrial enlarge*ment* is accentuated

associated pulmonary hypertension, the vessels had the same appearance as in uncomplicated atrial septal defect (Fig. 393).

The enlargement of the right atrium caused by the increased blood flow is a typical feature of atrial septal defect, but it may be difficult to establish roentgenologically. In the opinion of Bedford *et al.* (51), this is to be ascribed to the displacement of the tricuspid orifice to the left, which Roesler (562) has said to be a characteristic



Fig. 393 Atrial septal defect. Boy, aged 7 (C.H. 461121) see Figure 378, *J* and *K* Large left to right
greatly
thoracic
lated infundibulum Considerably increased vascularity both centrally and peripherally,
ment of right atrium, but not of left, aorta narrow

finding at autopsy. In our series, which consisted for the greater part of children, the atrial enlargement was seldom a dominant feature on the roentgenogram The dilatation involves both the appendage and the anterior and lateral parts of the atrium, it is seen as a prolongation and sometimes as a bulging of the right cardiac outline (Fig 394). The auricular appendage may be prominent. If the heart is shifted to the left, the right cardiac outline may have the normal appearance (Fig. 395) and the atrial enlargement can be visualized only in the oblique projection. The selective dilatation of the auricular appendage and of the part anterior to the crista supraterminalis, is presumably due to the greater distensibility of the structure in this part of the atrium. Anatomically, the posterior wall bears a greater resemblance to the left atrium, which is known to be less distensible than the right. Distinct dilatation of the posterior part of the right atrium as well was present in only two of our cases, in both, there were an extremely large shunt and pulmonary hypertension, and in one of them a complete A-V block was superadded. These observations confirm Schwedel's findings (583) with respect to the right development of dilatation of the right atrium

Enlargement of the right ventricle was marked in most cases It was lacking in all cases with a small shunt and inappreciable or no increase in heart volume, and was difficult to evaluate in some cases with a relatively large shunt The enlargement is

due to dilatation, which involves both the inflow and the outflow tract. On the roentgenogram, it is seen as an increase in contiguity of the ventricle to the thoracic wall (Fig. 392). The dilatation of the infundibulum is visualized best in the lateral projection Dilatation of the right ventricle, which is usually associated with slight shifting of the heart to the left, is often considerable The right ventricle usually forms the apex. In most cases, the apex merges with the dome of the diaphragm. The apex, however, may be slightly lifted and curved, without being upturned An upturned apex was observed in only three of our cases. The borderline between the right and the left ventricle may be marked by a notch at the apex (Fig 396). In our series, this notch was not seen as often as in ventricular septal defect.

Marked curvature of the anterior wall of the right ventricle is not one of the features of uncomplicated atrial septal defect. This change in the outline is usually seen in hypertrophy of the actual wall musculature. In our cases of atrial septal defect, it was observed only when there was a complicating rise in pressure in the ventricle, irrespective of whether it was caused by pulmonary hypertension or by pulmonary stenosis

In atrial septal defect, the aorta is narrow, depending on the reduced flow in the systemic circulation In one-fourth of our cases it was, however, of the normal width Irrespective of the size of the aortic arch, the segment outlined against the lung is smaller than its actual extent, owing to the dilatation of the pulmonary artery. The bulge made by the aortic arch in the esophagus is shallow In the oblique projection, the narrow ascending aorta cannot always be visualized without overlapping of the greatly dilated vessels around the hilum

The superior vena cava is generally poorly visualized, and in the frontal projection the right mediastinal outline often coincides with the paravertebral outline (Fig 392) This effect is caused by the decreased flow through the aorta and the vena cava and by the typical shifting of the heart to the left

Although the blood volume in the pulmonary circulation is increased with an atrial septal defect, the left atrium is not enlarged. This applies even when the shunt is large and has been observed by other authors (401). The absence of left atrial enlargement in the presence of a left to right shunt is crucial to the diagnosis and indicates that the shunt takes place above the atrioventricular plane. This feature, which in our experience is characteristic of both ostium primum and secundum defects, facilitates a differential diagnosis on roentgenologic examination from ventricular septal defect and patent ductus arteriosus. Moderate or inappreciable backward bulging of the atrium was seen in only six of our cases of atrial septal defect.

In our series, the heart volume was almost consistently increased, in several cases to more than twice the normal volume The largest hearts were observed in the presence of a septum primum defect. However, in the majority of these cases, enlargement was of the same degree as in the septum secundum defects Only in six cases with an extremely small shunt was it normal The successive enlargement of the heart during the first year of life in one of our cases is illustrated in Figure 397

Atrial septal defect complicated by raised pressure in the right ventricle.—In 10 of these cases (six children and three adults) *valvular pulmonary stenosis* or *infundibular stenosis* (one case) was present. In eight of them there was increased curvature of the anterior surface of the heart, indicating hypertrophy of the right ventricle (Fig 398). In other respects, the roentgenologic appearance was the same as in uncomplicated atrial septal defect, In the other two cases, all the features were identical The roentgenologic appearance and autopsy findings in a case of atrial septal defect with complicating mild infundibular stenosis are shown in Figure 399.

In cases with pulmonary hypertension and increased ...



Fig. 395.—Atrial septal defect. Man, aged 35 (W.M. 160106). The heart is shifted to the left Enlargement of the atrium is difficult to judge in the frontal view. There is considerable dilatation of the main trunk of the pulmonary artery, and both the aorta and superior vena cava are narrow



Fig. 396.—Atrial septal defect Boy, aged 9 (B.O. 430820) Borderline between right and left ventricles is seen as a shallow notch (arrows).



Fig 398 —Atrial septal defect and valvular pulmonary stenosis Girl, aged 4 (H N 480622) Right ventricle large and hypertrophic, with bulging of anterior surface, increased vascularity of lungs both centrally and peripherally, enlargement of right atrium aorta and superior vena cava narrow

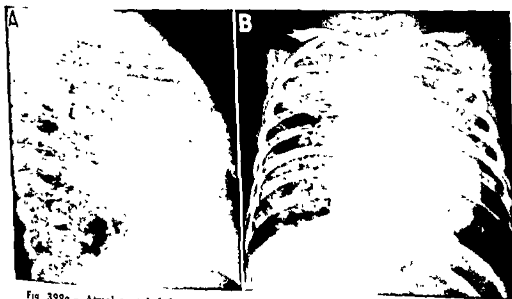


Fig 399a —Atrial septal defect and [illegible]
Boy, aged 5 months (S G-V 530830
great increase in vascularity of lung
of pulmonary artery

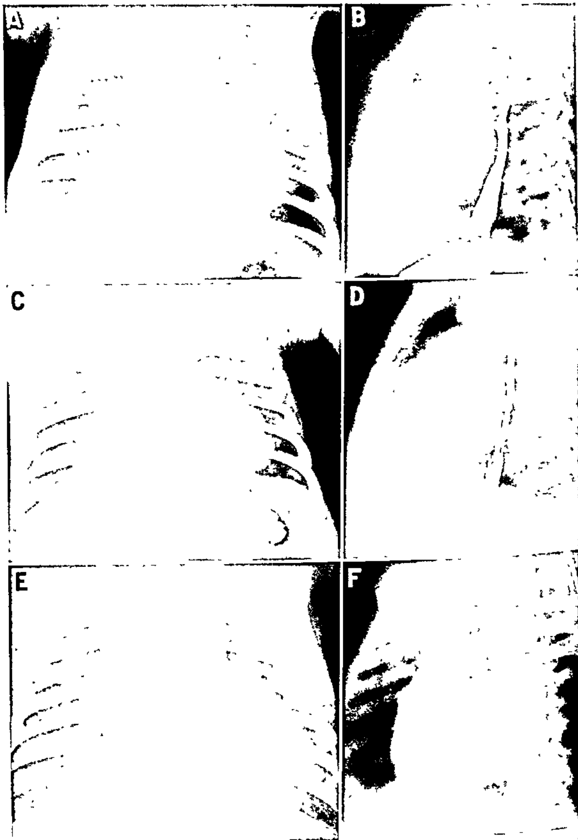


Fig. 397.—Atrial septal defect Boy (A T 520827) Size and configuration of the heart during infancy A and B, at 6 weeks of age, no increase in heart volume, outline normal C and D, at 3 months, slight enlargement of heart E and F, at 7 months, outline typical of atrial septal defect, moderate enlargement of heart, increased vascularity of lungs. Enlargement of right atrium difficult to determine



Fig 400 —Atrial septal defect and mitral stenosis (Lutembacher's syndrome) Girl, aged 7 (R.J 461030) As in uncomplicated atrial septal defect with a large left to right shunt, there are enlargement of the right ventricle and atrium, dilatation of the pulmonary artery, and greatly increased vascularity of the lungs, both centrally and peripherally. No enlargement of the left atrium Narrow aorta Pulmonary hypertension Anterior surface of the right ventricle is slightly bulging



Fig 399b.—Same case as in Figure 399a Great over-resorption of septum primum and ... and to right

... are somewhat short and thick Hypertrophy of trabeculae in the right ventricle, ... apart from hypertrophy of the actual wall muscles Appearance of left ventricle (*E*) is normal, apart from extensive network of trabeculae (see Fig 59, p. 58) White arrow in *D* points to mitral orifice. *CSV*, crista supraventricularis, *I*, infundibulum, *LAA*, left auricular appendage, *MV*, mitral valve, *OI*, ostium infundibuli, *PA*, pulmonary artery, *PB*, parietal band, *PM*, papillary muscle, *PSV*, pulmonary semilunar valve, *TV*, tricuspid valve, *VS*, ventricular septum.

right ventricle also presented the change in shape characteristic of hypertrophy. Both the main trunk of the pulmonary artery and its central and peripheral branches were dilated. The left atrium was not enlarged (Fig. 393).

An examination was made in one case of coincident *mitral stenosis (Lutembacher's syndrome)*, disclosed at autopsy. The roentgenogram and the fluoroscopic findings were typical of atrial septal defect. None of the features gave direct reason to assume the presence of mitral stenosis The left atrium was not dilated. The aorta was extremely narrow, whereas the pulmonary artery exhibited considerable dilatation (Fig 400)

Complicating *aortic stenosis* was present



Fig 400 —Atrial septal defect and ...
(R.J 461030) A...
enlargement of
increased vascu
...trum Narrow
bulging

in one case and could not be diagnosed roentgenologically.

The roentgenologic appearance in uncomplicated atrial septal defect is typical but not specific. The same roentgenologic features are present when there is a pulmonary *vein with an aberrant opening into the* right atrium, and a distinction between these two conditions can be made only by means of special investigations (see p. 444). With complete anomalous venous return the right atrium is, however, usually greatly dilated and the heart is grossly enlarged. In the majority of cases, it is not difficult to distinguish between a ventricular and an atrial septal defect When the latter is uncomplicated, dilatation of the left atrium is seldom found, whereas this is the rule in a ventricular septal defect with a left to right shunt. In atrial septal defect, the roentgenogram shows dilatation of the enlarged right ventricle, in ventricular septal defect, the curvature of its anterior surface caused by the hypertrophy is more conspicuous. The aorta is fairly often narrow in ventricular septal defect as well, and this feature is of very limited value as far as a differential diagnosis is concerned When atrial septal defect is complicated by raised ventricular pressure, the appearance is in several respects similar to that in ventricular septal defect with a mixed shunt.

No conclusive evidence has been brought forward of the possibility of diagnosing a *coincident mitral stenosis by means of* roentgenologic examination Both an aneurysmal dilatation of the pulmonary artery and an exceedingly narrow aorta may be found in uncomplicated atrial septal defect as well. If an enlarged left atrium is present, this is rather an argument against Lutembacher's syndrome, provided there is no heart failure.

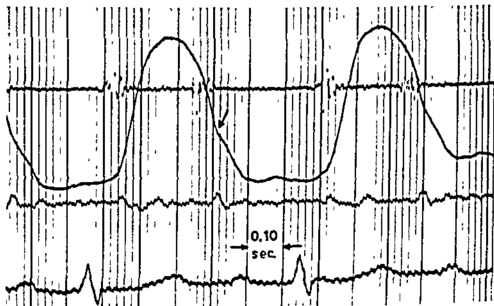


Fig. 401.—Electrokymogram of pulmonary artery in atrial septal defect. Girl, aged 7 (B L 460110) PCG over apex Frequency channel, 20 cps Time marking, 0 10 sec between thick lines Onset of rise is rounded Gradient of upswing is normal Incisura (arrow) is poorly defined, since the dicrotic wave is absent Pressure: in RV 35/4 mm Hg, PA 33/4

ELECTROKYMOGRAPHY

The electrokymograms recorded over the pulmonary artery in atrial septal defect with a left to right shunt often show typical alterations. The tracings from the aorta atria, and ventricles, on the contrary, usually show no distortion.

Electrokymograms were recorded in 30 cases, neither right ventricular failure nor pulmonary hypertension was present in any of them An incomplete right bundle-

ranch block was present in 29 cases and a omplete block in one

Abnormal tracings from the pulmonary rtery are demonstrated in Figures 401–05. The characteristic features were the

sound (Fig 404). The slope of the upstroke was normal, and the rise occurred at a normal interval after Q-R. The onset was sometimes round and therefore not always exactly identifiable

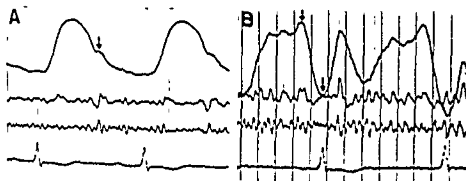


Fig 402.—Fl...

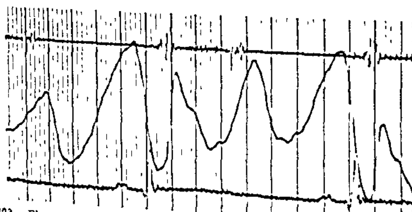


Fig 403 —Fl...

broad and strikingly *round* summit and the late transit into the diastolic phase The positions of the incisura and the dicrotic wave on the descending limb varied, but they were never high up The dicrotic wave was usually low The incisura coincided with the pulmonary component of the second

... an impediment to atrial emptying

Tracings with this appearance were recorded in 11 of the 27 cases in which there was *no* associated pulmonary stenosis. In the remaining 16 they were normal The alterations described in the electrokymogram of the pulmonary artery presumably imply that, during the prolonged reduced

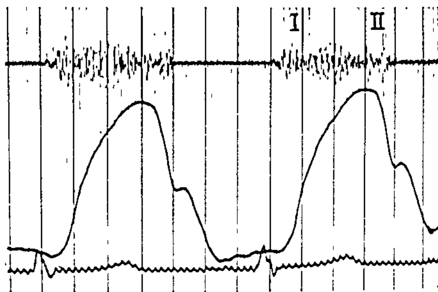


Fig. 404.—Electrokymogram of pulmonary artery in atrial septal defect. Boy, aged 11 (B O 430820). PCG over pulmonary area, *I*, 1st sound, *II*, 2nd sound, split. In comparison with preceding cases, the appearance of the electrokymogram is somewhat modified, as maximal dilatation of the pulmonary artery occurs at the end of systole. Protodiastolic phase, 0 06 sec. Dicrotic wave is low. Incisura coincides with the second component of the 2nd pulmonary sound. Pressure: in RV 35/2 mm Hg, PA 35/7.

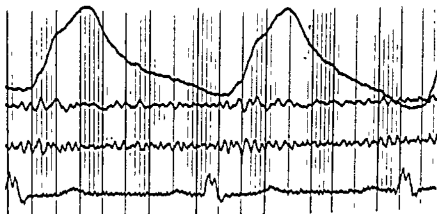


Fig. 405.—Electrokymogram of pulmonary artery in atrial septal defect. Man, aged 37 (O M 160106). Complete right bundle-branch block Frequency channels, 10 and 20 cps. Appearance of this electrokymogram differs from the others An inflection is present in the prolonged upstroke Protodiastolic phase cannot be defined, and incisura and dicrotic wave are reduced Pressure in RV 21/0 mm Hg, PA 20/12 Extremely large left to right shunt

ejection, this artery sustains its distention and that its collapse during diastole occurs more continuously than normally, without any marked rebound on closure of the valve. This is probably caused by the increased stroke volume in the right ventricle and the decreased resistance in the pulmonary circulation, with a resulting rise in the pulse pressure. There was, however, no simple correlation with the findings in the electrokymogram, since normal tracings were sometimes recorded in cases with a greatly increased flow and raised pulse pressure

With associated pulmonary stenosis, electrokymographic alterations typical of valvu-

lar stenosis were found in two of three cases.

Abnormal electrokymograms from the right atrium were recorded in four cases (Figs 335 and 403). Their appearance was that in the presence of an impediment to atrial emptying Ödman (516) recorded such abnormal tracings over the right atrium in the majority of his series of 32 patients, chiefly adults, with atrial septal defect. This is probably to be ascribed to the fact that hemodynamic changes were more advanced than in our series

In the tracings from the ventricles, the diastolic limb showed such large variations in the individual cases that no conclusions regarding the filling conditions in these chambers are warranted on the basis of its shape.

CARDIAC CATHETERIZATION

The clinical features had long given reason for suspicion that the blood is shunted from left to right through the atrial septal defect (562) With the introduction of cardiac catheterization, it became *possible to* confirm this supposition (85, 189)

INTERATRIAL PRESSURE RELATIONS — Pressure recordings from both the right and the left atrium were made for the first time by Cournand *et al* (165) in three cases of atrial septal defect in children In one of these cases, however, there was coincident pulmonary stenosis, with a systolic pressure of 59 mm Hg in the right ventricle The right ventricular pressure was also raised *in the* other two cases The mean pressure in the left atrium was 1 6 to 4 mm higher th

[illegible] atrial systole (the *a* wave) was more marked in the left atrium (*b*) A positive notch, corresponding to the beginning of ventricular systole, was present in the right atrial curve It was lacking in the left (*c*) The fall in pressure associated with the displacement of the atrioventricular plane toward the apex during ventricular systole was less marked in the right atrium than in the left. (*d*) The rise in pressure during the ventricular ejection phase was considerably greater in the left atrium. (*e*) The fall in pressure during the phase of rapid ventricular filling was more marked in the left atrium. The pressure in the left atrium was higher during the whole of the cardiac cycle, with the exception of a short period corresponding to point (*c*). This was stated to be dependent on the fact that upward and downward movement of the ventricular base was more marked on the left side, a fact pointed out earlier by Wiggers For this reason, a right to left shunt could also arise. Soulié (620) stated that the pressure in the right atrium attained a maximum more rapidly during ventricular *systole*, with a right to left shunt as a result. Calazel *et al* (127) found that, with an uncomplicated atrial septal defect, the left atrial pressure exceeded the right during most of the cardiac cycle. The right atrial pressure was higher only at the beginning of auricular filling. In these cases, a small right to left shunt was present in addition to the dominant left to right shunt; it was considered to be caused by the reversal in the pressure gradient at the beginning of auricular *filling*

Pressure recordings from the left and right atria have seldom been made in healthy subjects We had the opportunity of making such a recording in a 6-year-old boy (U W *470212*), he had never shown any symptoms of heart disease, and the electrocardiogram and roentgenologic ap*pearance of the heart* were normal He was admitted to our clinic on account of a systolic murmur, *which* we nevertheless interpreted as a physiologic murmur. Cardiac catheterization showed normal pressure in the right ventricle and pulmonary artery and no shunts. The catheter passed through a *patent* foramen ovale The pressure recordings from the atria (Fig. 406, *A*) exhibited, on the whole, the same features as those described by Cournand *et al* [illegible]

[illegible] decrease in pressure [illegible]

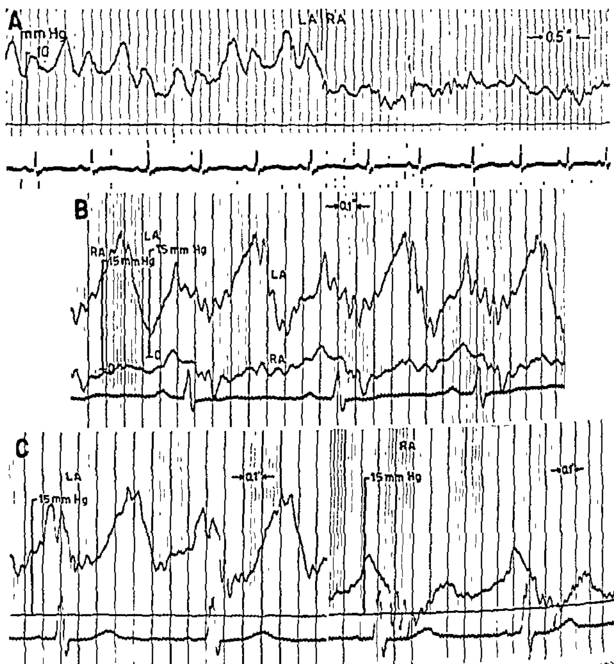


Fig. 406.—Pressure recordings from right and left atria. *A*, healthy boy, aged 6 (U W. 470212) with a physiologic, systolic murmur Withdrawal curve on passage of catheter from the *left to* the right atrium through a patent foramen ovale. *B*, girl, aged 7 (B O. 461003). Simultaneous recording with one catheter in each atrium Extremely small patent ductus arteriosus, normal pressure in right ventricle, patent foramen ovale *C*, boy, aged 7 (B S 460607). Coarctation of aorta and patent foramen ovale

sponding to the movement of the ventricle downward toward the apex was almost equally marked in both atria. (3) Pressure in the left atrium was higher than in the right during the whole cardiac cycle. The greatest difference in pressure was recorded slightly before the opening of the atrioventricular valves, and the next greatest during atrial systole The respiratory variations in the pressure were more conspicuous in the left atrium, as had been shown earlier in the dog by Brecher and Opdyke (89).

Pressure recordings from both atria were made simultaneously with two catheters in one case in which the catheter passed through a patent foramen ovale, no interatrial shunt was present and pressure in both ventricles was normal (Fig 406, *B*) The patient was a 7-year-old girl (B.O 461003) with a small *patent ductus* The shunt was extremely small (0 9 volume per cent difference between the right *ventricle* and the pulmonary artery) There was good reason to suppose that this small shunt would have little influence on the pressure in the *left atrium* The pressure curve was, in fact, similar to that in the normal case The most salient difference between the pressu... *a* wav... right ... in the left atrium the *v* wave was higher ..., whereas Figure 406, *C*, shows the recordings from both atria in a case of uncomplicated coarctation of the aorta with a patent foramen ovale (a 7-year-old boy, B.S. 460607). The blood pressure in the upper half of the body was slightly raised (140/90 mm Hg), and there was only slight enlargement of the left atrium and left ventricle. The atrial pressure curves did not differ from those in the normal case.

In atrial septal defect, the pressure remains higher in the left atrium than in the right (288, 441), but some interatrial equilibration of the pressure takes place. If the defect is extremely large, the same pressure is recorded in all four chambers of the heart during the end of diastole (187) Figure 407 shows the pressure recording from both atria in a patient who had an uncomplicated but large atrial septal defect, verified at operation. When the defect *is small*, the course of the pressure in the right atrium remains at normal levels (Fig. 408)

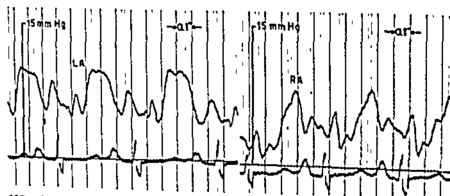


Fig. 407.—Pressure recordings from right and left atria in large, uncomplicated atrial septal defect Girl, aged 8 (B L 460110) Pressure curves are fairly similar Right atrium has an abnormally high *v* wave

The normal interatrial pressure relation may be entirely altered when there are malformations of the atrioventricular valves (see p 731) or in conditions which cause increased right ventricular pressure. The atrial pressure in pulmonary stenosis has been discussed on page 186, and Figure 182 shows the pressure recordings from both atria in a case of severe pulmonary stenosis. The tall *a* wave in the right atrium is considerably higher than the *a* wave in the left atrium, whereas the *v* wave remains *higher in the latter*. The same change in atrial

*pressure is found in right ventricular hyper*trophy caused by a large ventricular septal defect and great resistance in the pulmonary circulation. Figure 409 shows simultaneous pressure recordings from both atria in such a case (15-year-old girl, G.G. 390314); the resistance in the pulmonary circulation was so great that a right to left interventricular shunt had developed, and the left to right shunt was extremely small. The catheter had passed through a patent, non-functioning foramen ovale. The pressure was almost identical in the two atria. In this case, the blood flow was greater *This is because of the* greater distensibility of the right atrium. In the left atrium, which is less distensible, the *v* wave is higher. With the opening of the mitral valve, an unusually rapid fall in pressure *occurs in the left atrium* and the *a* wave is low. Both of these features indicate low resistance to the flow to the left ventricle, which has become more thin-walled and distensible. The respiratory variations are more marked in the left atrium (see p. 439). The interatrial shunt passed alternately in both directions, largely due to the respiration. Since the ductus arteriosus was

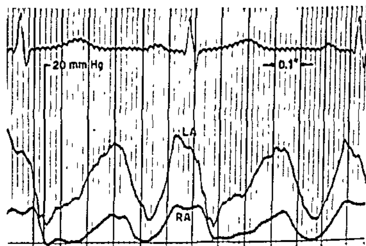


Fig. 408.—Simultaneous pressure recordings from both atria in a case of small atrial septal defect Girl, aged 8 (G H 460502) Pressure in the right atrium is the same as in normal subjects

through the right atrium than through the left.

In this connection, it is of interest to consider the change in the atrial pressure when *the pressure in the two ventricles is re*versed. We performed cardiac catheterization in one case of transposition of the great vessels with an atrial septal defect and an intact ventricular septum Pressure tracings were obtained from both atria and both ventricles. Systolic pressure was 19 mm Hg in the left ventricle and 78 mm in the right. Figure 694 (p. 788) shows the pressure recordings from the atria It is seen that the *a* wave in the right atrium is high, as an expression of the increased pressure required to fill the hypertrophied right ventricle. The *v* wave, in contrast, is not abnormally high, despite the large flow not patent (verified on angiocardiography) the mixed shunt was essential for life.

In uncomplicated atrial septal defect, we have consistently found higher pressure in *the left atrium* throughout the cardiac cycle Our series, however, consisted exclusively of children, and none of them had heart failure at the time when catheterization was performed. In cases with pulmonary hypertension, especially when right ventricular failure develops, the atrial pressure changes and a right to left shunt can be demonstrated (152, 187, 351, 451, 642)

Selzer and Lewis (594) assembled the cases of atrial septal defect with chronic cyanosis reported *in the literature*. Cardiac catheterization was not, however, performed in these cases. As in a later publication by Selzer (590), it is stressed that a

ght to left interatrial shunt may occur uring a certain phase of the cardiac cycle, ven with no change in the pressure rela- ons In this event, a venous flow—particu- rly from the inferior vena cava—could asily pass through the defect, although the

cava, there are great anatomic possibilities of a shunt from the inferior vena cava to the left atrium. In these cases as well, we ... the 32 ...fect in-

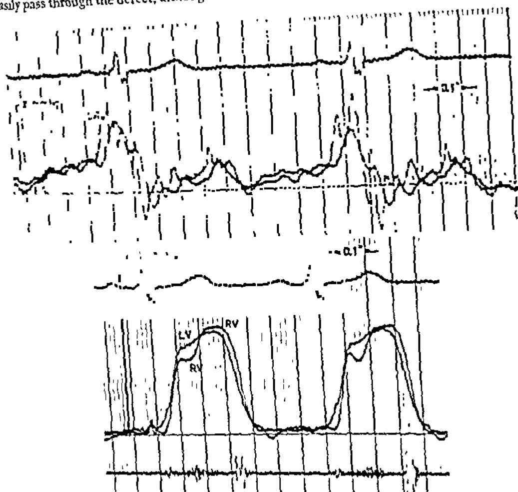


Fig 409.—Pressure recordings from atria and ventricles ...
Girl, aged 15 (G G 390 ...
cies and great resistanc...
both atria LA and RA, ...

pressure need not be lower in the left atrium.

A small right to left interatrial shunt was present in three of our cases, one with an ostium primum defect and two complicated by pulmonary hypertension If the defect lies far down and posteriorly in the septum, at the orifice of the ...

vestigated at the Department of Clinic... Physiology, Karolinska Sjukhuset, had ... significant right to left shunt at rest, but heavy work load produced a right to l... shunt in two of them. One had pulmona... hypertension, and the other a defect si... ated at the orifice of the inferior vena ca...

er injection of dye, Swan *et al.* (637, 641, 642) were able to demonstrate a small right to left shunt causing no significant decrease in arterial oxygen saturation. As a rule, the shunt was most easily recorded on injection of dye into the inferior vena cava, but in high-lying defects combined with anomalous connection of right pulmonary veins to superior vena cava, the shunt was best recorded by injection into the superior vena cava. A right to left shunt demonstrable by gas analysis was, however, found in only one case with high right ventricular pressure.

The results of angiocardiographic examinations have also been brought forward as evidence of a mixed shunt in atrial septal defect. Lind and Wegelius (438) demonstrated passage of the contrast medium from the right to the left atrium when the injection was made into the inferior vena cava. It is not evident from their publication whether the patients in question had an uncomplicated atrial septal defect or whether there was coincident right ventricular hypertrophy or tricuspid valve disease. Pulmonary stenosis was present in two of the cases in which Friedlich *et al* (264) demonstrated a right to left shunt on angiocardiography. In two cases of uncomplicated atrial septal defect we injected the contrast medium into the inferior vena cava, but were unable to observe any passage of the contrast medium from the right to the left atrium, we therefore abandoned this method (see p. 450).

In atrial septal defect, a decrease in the arterial oxygen saturation is not proof of a right to left shunt When there is an extremely large left to right shunt, there is sometimes reduced oxygenation of the blood in the lungs Many writers have repudiated this possibility, whereas others have found evidence in support of it This question has been discussed on page 372, in connection with ventricular septal defect Wood (714) found decreased oxygen saturation in samples from the pulmonary veins in some cases of atrial septal defect and stated that the rapid rate of flow through the capillaries of the lungs might be responsible. In 64 of our cases the catheter passed through the defect, and in four of them the pulmonary venous samples were unsaturated; the figures were 86, 87, 90, and 88 per cent, respectively.

To sum up, it may be stated that the occurrence of a left to right shunt in atrial septal defect is unquestionable. Some authors (127, 590, 637) have found that a right to left shunt may be present in addition. Our experience with a series of children is that a right to left shunt of sufficient size to be demonstrable by gas analysis is lacking in the uncomplicated cases

Little (440) has shown in experiments on dogs that the distensibility of the right atrium is considerably greater than that of the left. The anatomy of the two atria indicates that this also applies in man (see p 75). Hull (351) pointed out that the inflow to the left ventricle offers greater resistance than the inflow to the right. This is because the left ventricle has a thicker wall, the mitral orifice is smaller than the tricuspid orifice, and the inflow tract of the left ventricle is narrower and more elongated than that of the right. Dow and Dexter (205) and McCord *et al* (451) have stressed that the pressure-volume relationship of the right ventricle is decisive for the direction of the flow. The thin-walled right ventricle is more distensible and can undergo a greater increase in volume than the left, with a corresponding diastolic pressure If the defect is very large, functionally a common atrium, the same pressure is obtained during end-diastole in both atria and both ventricles (187), but the diastolic volume is then considerably greater in the right ventricle than in the left. This relationship is altered in right ventricular hypertrophy

The blood flow through the defect is dependent on the size of the defect and the pressure difference between the left and right atrium. Although this pressure difference is small, it is nevertheless present in both systole and diastole, and the shunt may therefore be considerable The defect must, however, be at least 1 cm in diameter

an adult for any significant shunt to se.

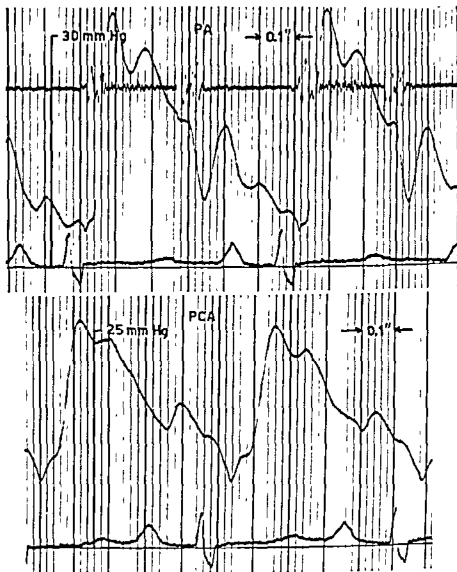
PULMONARY CAPILLARY ARTERIAL PRESSURE PULSE (PCA PRESSURE).—Wood (713) emphasized that if the catheter is advanced so far into a pulmonary vein that the vessel is blocked, the arterial pulse curve may resemble a pressure curve from the pulmonary artery. Weissel *et al.* (692, 693) gave a more detailed description of this phenomenon. They found, in a series of 12 cases, that PCA pressure did not exceed 33 mm Hg even when pressure in the pulmonary artery was considerably raised. The pressure is damped in the small arteries, which are consistently narrowed.

The PCA pressure was recorded in 60 of our cases of atrial septal defect, and in all except four the pulmonary artery pressure was also recorded. Tracings of the PCA pressure were made in an additional 67 patients with various kinds of heart disease. Among them were patients with normal pressure in the pulmonary artery, with pulmonary hypertension and with pulmonary stenosis, the pulmonary artery pressure was recorded in 59 of them. Good agreement was found between the pulmonary artery pressure and the PCA pressure (Figs. 410 and 411). In 14 cases, marked pulmonary hypertension was present. The PCA pressure was damped (Fig. 412) in these cases, and the highest mean pressure recorded was 40 mm Hg.

Wood (714) obtained a typical arterial pulse curve only in the presence of an atrial septal defect in which the flow was large and the resistance in the pulmonary circulation low. In other cases, in which the catheter penetrated a patent foramen ovale, the curve was damped. In our experience, an undamped arterial pulse curve is obtained even in cases without a shunt and with normal pressure in the pulmonary artery (see Fig. 413).

CATHETERIZATION AS AID IN DIAGNOSIS OF ATRIAL SEPTAL DEFECT.—In cardiac catheterization, the criterion of an atrial septal defect is greater oxygen saturation of samples from the right atrium than from the venae cavae. This cannot, however, be accepted as proof, since the same result is obtained in other malformations, i.e., (1) a pulmonary vein opening into the right atrium (possibly through the coronary sinus), (2) a ventricular septal defect with tricuspid incompetence, (3) a defect between the left atrium and the coronary sinus, (4) a defect between the left ventricle and the right atrium (216, 250), and (5) a communication between the aorta and the right atrium. The last-mentioned anomaly is extremely rare and results in entirely different clinical features, including a loud continuous murmur.

The blood entering the right atrium through the superior and inferior venae cavae and through the coronary sinus differs greatly with respect to the oxygen saturation. Because the blood is poorly mixed in the atrium, gas analysis provides no definite information about the shunt. During ventricular diastole, part of the flow from the atrial septal defect passes directly into the ventricle. Consequently, the oxygen saturation of samples from the right ventricle is often found to be appreciably higher than that of samples from the atrium, which would seem to be an indication of ventricular septal defect. It has been considered that the oxygen content of the superior vena cava and of the right atrium must differ by at least 2 volumes per cent to permit a diagnosis of atrial septal defect (292, 314). Greater certainty can be insured if a large number of samples from both venae cavae and from different parts of the atrium are analyzed. The volume of the shunt can best be estimated by calculating the difference between the oxygen saturation of the blood from both venae cavae and from the pulmonary artery, in which the blood is effectively mixed. However, it is then necessary to demonstrate the presence of a communication between the atria by some other method and at the same time to rule out a left to right shunt to the right ventricle or to the pulmonary artery. Both a ventricular septal defect and a patent ductus can be ruled out on the basis of the physical find-



[illegible] right shunt and nor- [illegible] de in the pulmonary [illegible] volume and low pe- [illegible] 08 sec after the PA pressure.

electrocardiography, phonocardiography or angiocardiography.

The most definite evidence of an anomalous communication between different chambers of the heart or the vascular system is the passage of a catheter through it If the catheter passes from the right to the left atrium, a non-functioning but patent foramen ovale may, however, also be present. This happens very easily if the catheter is introduced into the atrium through the inferior vena cava. If a left to right shunt can be demonstrated at the same time, it may be due to the opening of a pulmonary vein into the right atrium Consequently, the passage of the catheter from the right to the left atrium is not, in itself, of any diagnostic value. On the other hand, catheterization of the left atrium and pulmonary vein provides valuable information about the oxygen saturation of the pulmonary venous blood and the presence of a right to left interatrial shunt. By means of angiocardiography with injection into the left atrium, it can be established whether the catheter has passed through a patent fora-

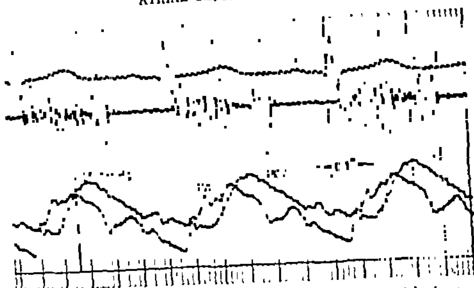


Fig. 411.—PA and PCA pressures in atrial septal defect with small left to right shunt and normal pressure in pulmonary artery Girl, aged 8 (G H 460502) Pulse amplitude is not as large as in Figure 410 Time interval between the pressures is 0 11 sec

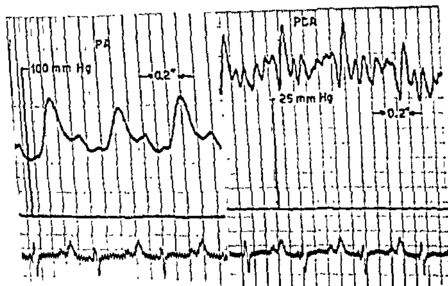


Fig 412 —PA and PCA pressures in Lutembacher's syndrome with pulmonary hypertension Girl, aged 6 (R.J 461030) PCA pressure is not typical of an arterial pulse curve

men ovale or a true atrial septal defect (p 454).

We have earlier, with Bjork and Crafoord (64), described a simple method for distinguishing between a true defect and a patent foramen ovale which is covered by the valve A catheter, provided with a balloon on the tip, after Hanson (317), is introduced into the left atrium and filled with contrast medium (30 per cent Diodrast or Urokon) so that it can barely be drawn back into the right atrium (Fig. 414). This gives an idea of the size of the defect. If the catheter has passed through a foramen

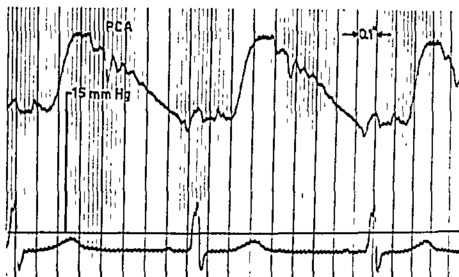


Fig. 413.—PCA pressure in coarctation of the aorta Boy, aged 7 (B S. 460607). Catheter passed through a patent foramen ovale. No shunts were present, and pressure in the pulmonary artery was normal PCA pressure exhibits a typical, undamped arterial pulse curve.



Fig 414.—Atrial septal defect Boy, aged 3 (B S 500913) The balloon is so distended that it can barely be drawn back from the left atrium into the right. If the catheter passes through a patent foramen ovale covered by the valve, it must be entirely emptied before it can be passed from the left to the right atrium

ovale entirely covered by the valve, the balloon must be emptied if it is to be withdrawn into the right atrium. The septum may occasionally be fenestrated, and a mistake may be made if the catheter has penetrated a small hole instead of the large defect Varnauskas and Werko (670) have described a catheter provided with two balloons which permits closure of the defect. With another catheter, the right side of the heart can be catheterized again, and the effect on the oxygen content and pressure can be recorded

Some idea of the position of the defect vein has advanced via a septal defect and the left atrium or has passed directly into the pulmonary vein from the right atrium The existence of a venous connection can be established with certainty on the basis of the position of the catheter only if the pulmonary vein opens into the superior vena cava above the right atrium A foramen ovale defect is most easily penetrated if the catheter is introduced through a saphenous vein, whereas a sinus venosus defect is best catheterized through an arm vein When evaluating the position of a defect on the basis of the course of the catheter, it must be borne in mind that all types of atrial septal defect may be combined with a patent foramen ovale. Thus, when an ostium primum defect is present, the catheter may advance through a patent foramen ovale and the position of the defect be misinterpreted

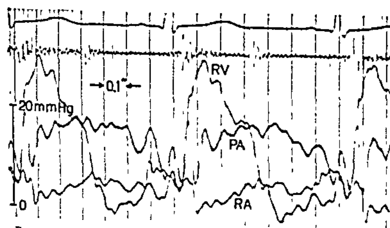


Fig 415 —Pressure records from right atrium (*RA*), right ventricle (*RV*), and pulmonary artery (*PA*) in atrial septal defect Man, aged 29 (Case 53/56) The ratio of pulmonary to systemic flow was 3 6 1. At the end of diastole, the pressures in right atrium, right ventricle, and pulmonary artery are on the same level

can be obtained by observing on the fluoroscopy screen how the catheter passes through the defect (70, 293, 642, 681). In an ostium primum defect, the catheter advances far down toward the atrioventricular plane A foramen ovale defect lies between this plane and the orifice of the superior vena cava, in the anteroposterior projection, it lies somewhat to the right of the midline When a sinus venosus defect is present, the catheter passes through it high up, at the orifice of the superior vena cava Moreover, in such cases there is generally anomalous drainage of pulmonary veins It may, however, be difficult or impossible to determine on fluoroscopy whether a catheter lying in a pulmonary

The dye-dilution curve technique may provide valuable information about the position of the defect and opening of pulmonary veins (640, 642, 644, 681, 691).

In atrial septal defect, the resistance in the pulmonary vascular bed is low. Despite a pulmonary flow of up to 20 liters per minute in adults, the pressure in the pulmonary artery is generally normal (366, 714) If the shunt is large, the pulse amplitude is

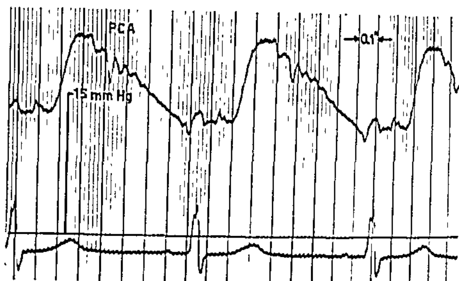


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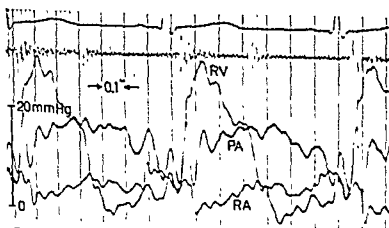


Fig 415 —Pressure record- fr... artery (PA ... temic flow ... pulmonary ... ind pulmonary ... monary to sys- ... ventricle, and

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increased, with a somewhat high systolic pressure and a diastolic pressure close to the end-diastolic pressure in the right ventricle (Fig. 415). Thus the mean pressure is normal and the resistance lower than in healthy individuals at rest. Normally, the pressure does not rise during exertion until the blood flow has increased more than threefold (162). In atrial septal defect, on the other hand, a rise in pressure has been recorded during exertion (333), but in a group of adult patients we found a rise usually no more marked than in healthy subjects (366).

Severe pulmonary hypertension is considerably more uncommon than in ventricular septal defect and patent ductus arteriosus. Nor are the changes in the small pulmonary arteries characteristic of pulmonary hypertension (see p. 339) seen as often in atrial septal defect (216, 219, 323). An extremely large ventricular septal defect or a wide patent ductus results in the same pressure in the systemic and in the pulmonary circulation. Only by means of increased resistance in the pulmonary circulation can the shunt be limited, so that an adequate systemic circulation is maintained. In atrial septal defect, on the contrary, the shunt is limited by the diastolic filling of the right ventricle. It is evident from Figure 416 that in the cases with the largest shunt, the pulmonary artery pressure was not remarkably high Pulmonary hypertension was present in only three cases. The shunt was fairly small in only one of them, indicating that the resistance was high

The cause of the high resistance in the pulmonary circulation in some cases is not known. It is present more often in adults than in children (34, 186, 187, 219, 380, 502, 691). Occasional cases with pulmonary hypertension in childhood have nevertheless been reported in large series (139, 196, 691). Wood (714) has stated that in the few cases of atrial septal defect in which pulmonary hypertension is present, the pulmonary changes have not developed as a result of the large pulmonary flow but have existed since very early childhood. Of 32 adult patients investigated at the Department of Clinical Physiology, Karolinska Sjukhuset, only one had moderately raised pulmonary vascular resistance, and, judging by the history, it had been present since early childhood.

The fact that the incidence of pulmonary hypertension has often been found to be higher in adults than in children is not proof that the lesions have been acquired in adulthood, as a result of the large pulmonary flow Naturally, the incidence of various types of heart disease investigated at our laboratories does not represent the true incidence It is probable that the proportion of children and adults is not the same, but it is not known which category is predominant Adult patients with pulmonary hypertension may be over-represented A small atrial septal defect causes no symp-

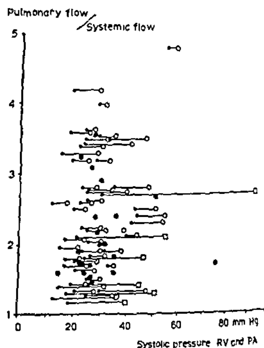


Fig. 416.—Atrial septal defect; relation of size of the left to right shunt (expressed as ratio of pulmonary to systemic flow) to systolic pressure in the right ventricle (open circles) and pulmonary artery (dots). In cases with pulmonary artery pressure lower than pressure in the right ventricle, a line denotes the pressure gradient. When anatomic pulmonary stenosis is present, the right ventricular pressure is denoted by a square A filled circle denotes no pressure gradient.

toms even in adults, and the murmur is heard so faintly through a thick thoracic wall that heart disease is not necessarily detected, even if medical advice is sought. Children far more often than adults undergo medical examination, either at health check-ups or in connection with respiratory tract infections. The murmur is more ... nowadays

symptoms, and patients with pulmonary hypertension belong to this category. The question of the causation of pulmonary hypertension cannot be answered with any degree of certainty until the development has been followed in a large series of cases, from early childhood to maturity, by means of repeated examinations

Pulmonary hypertension is considerably more common in persistent ostium atrioventriculare commune, in view of the direct interventricular communication. Other types of atrial septal defect as well may be combined with ventricular septal defect and pulmonary hypertension

A small pressure gradient between the right ventricle and the pulmonary artery may occur even when there is no stenotic malformation in the outflow tract of the ventricle In the presence of a large left to right shunt, the rate of flow through the pulmonary orifice is so high that the pressure of velocity is responsible for an apparent pressure gradient (364)

True pulmonary stenosis was present in seven cases, valvular stenosis in six and infundibular stenosis in one In all these cases there was a left to right shunt only

Complicating mitral stenosis (Lutembacher's syndrome) was present in one case As already mentioned, it was not possible to make this diagnosis on the basis of the physical findings or the roentgenologic examination Nor did the pressure recording from the left atrium show a curve typical of mitral stenosis, this is not surprising in view of the drainage of the left atrium through the defect Mean pressure in the left atrium was 10 mm Hg An extremely large left to right shunt was present, and pressure in the pulmonary artery was raised (76/41 mm Hg). An atrial septal defect 2 5 cm in diameter and a mitral orifice about 7 mm wide were found at operation. Thus, it is exceedingly difficult to diagnose Lutembacher's syndrome clinically, and the diagnosis is, in fact, usually made at autopsy. Demonstration of a diastolic pressure gradient between left atrium and ventricle is of diagnostic value, but in this case the left ventricular pressure could not be recorded

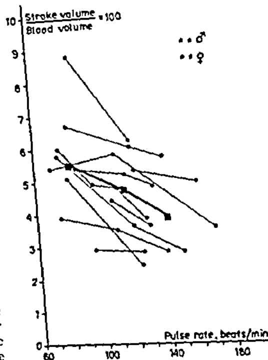


Fig 417.—Atrial septal defect, change in stroke volume of right ventricle with rising pulse rate during exercise The size of the stroke volume in the individual cases can be compared, since it is expressed as a percentage of the blood volume In healthy subjects at ...

mean values in all cases (From Jonsson et al., Acta med scandinav 159.275, 1957)

HEMODYNAMICS DURING EXERCISE

The effect of heavy work on the hemodynamics in atrial septal defect has bee

studied at the Department of Clinical Physiology, Karolinska Sjukhuset (366). It was found that the pressure in the right ventricle often does not rise more than in healthy subjects during exercise, but that the stroke volume of the ventricle is considerably reduced (Fig. 417). This implies that the increase in flow through the defect is not as great as the increase in flow in the systemic circuit, and in some cases it is even possible to demonstrate an absolute decrease in the shunt. Thus the load on the right ventricle does not seem to increase markedly during exercise. When the left to right shunt lies on the atrial level, the stroke volume of the left ventricle during exercise cannot be determined with the Fick principle. In patients with a small or moderately large defect, it seems to be normal, since these patients have a normal physical working capacity. In patients with a very large defect and low physical working capacity it is, on the contrary, low. This situation is also indicated by the anatomic conditions in these patients, i.e., a small left ventricle and narrow aorta.

ANGIOCARDIOGRAPHY

The diagnosis of atrial septal defect is usually well substantiated by the clinical and roentgenologic findings. Notwithstanding, this does not exclude the possibility that a pulmonary vein with an aberrant opening into the right atrium may actually be responsible for the changes, either wholly or partly. The effect on the hemodynamics is, in fact, the same in both of these conditions. A detailed analysis of the anatomic basis from this point of view can seldom be made at the roentgenologic examination.

Nowadays, an atrial septal defect is amenable to surgical treatment. A prerequisite for an intervention of this nature is that the existence of the defect has been convincingly demonstrated by means of one or other of the examinations. Thus, from the point of view of the surgeon, it is desirable for the presence of the defect to be established *anatomically* as well. Moreover, its size and position should be determined if possible.

We have already stressed that cardiac catheterization affords only limited possibilities of giving anatomic proof of the existence of an atrial septal defect, unless a balloon catheter is used. With respect to the possibility of demonstrating, in addition, the presence of pulmonary veins with anomalous openings, the reader is referred to page 522.

Many workers have endeavored to visualize the defect by means of angiocardiography. In our opinion, the results of examination with intravenous injection of the contrast medium do not warrant further attempts with this technique. This is because direct demonstration of the septal defect (or defects) presupposes a passage of the contrast medium from the right atrium to the left. In uncomplicated atrial septal defect, the flow—at least in our experience (see p. 442)—is directed from the left to the right atrium during the greater part of the cardiac cycle. Consequently, passage of the contrast medium in the opposite direction can be brought about only by reversing the shunt. The rise in pressure in the right atrium caused by rapid intravenous injection does not, as a rule, seem to suffice for this purpose. This applies, in our experience, even when the injection is made selectively through a catheter with the tip placed in the right atrium, immediately beside the septum and pointing to the defect (Fig. 418). It is true that the catheter recoils somewhat from the septum during the injection, but this is presumably only a minor cause of the lack of the desired effect.

When the contrast medium is injected intravenously or into the right atrium, an abnormal inflow into the atrium is sometimes disclosed by a local mixing defect. Interpretation of the implication of local filling defects in the right atrium is made more difficult by the fact that, even normally, blood flows into the right atrium from three directions, namely, both caval veins and the coronary sinus. A dilution effect can therefore be evaluated as path-

ologic only if it is at a typical site and appears when the atrium is well opacified. Under these conditions, Figley *et al* (254) observed such a pathologic mixing defect in three of 13 cases of uncomplicated atrial septal defect

In complicated atrial septal defect, which is associated with a marked rise in pressure in the right ventricle, the defect can sometimes be visualized irrespective of whether the contrast medium is injected intravenously or selectively into the right atrium The increased resistance in the right ventricle causes such changes in the interatrial pressure relations that passage of the medium to the left atrium takes place.

In uncomplicated atrial septal defect, the recirculation of the contrast medium to the right atrium is an expression of the left to right shunt occurring in this condition As in ventricular septal defect, the practical value of demonstrating recirculation appears to be insignificant Moreover, this observation must be interpreted with caution (204). Owing to dilution by the blood, particularly during passage through the pulmonary circulation, the density of the contrast medium is usually greatly reduced. Considerable difficulties may therefore be encountered in determining whether it is actually present in the right atrium during recirculation. Soloff and Zatuchni (618) nevertheless attached great importance to this reopacification of the right atrium and claimed to have demonstrated that it is possible, with this method, to distinguish between septum secundum and primum defects They stated that in the former type the posterior border of the right atrium is hazy and irregular, owing to local dilution by blood from the shunt, partial opacification of the left atrium occurring simultaneously In septum primum defects, on the contrary, they found poor definition of the inferior portion of the dorsal border of the right atrium, as well as opacification of the left ventricle as soon as the right atrium became filled with contrast medium In



Fig. 418.—Atrial septal defect of foramen ovale type Boy, aged 6 (S P 470214) Tip of the catheter is placed in the right atrium, beside the septal defect and directed toward it The jet of contrast medium deviates and is collected by the powerful blood flow through the septal defect.

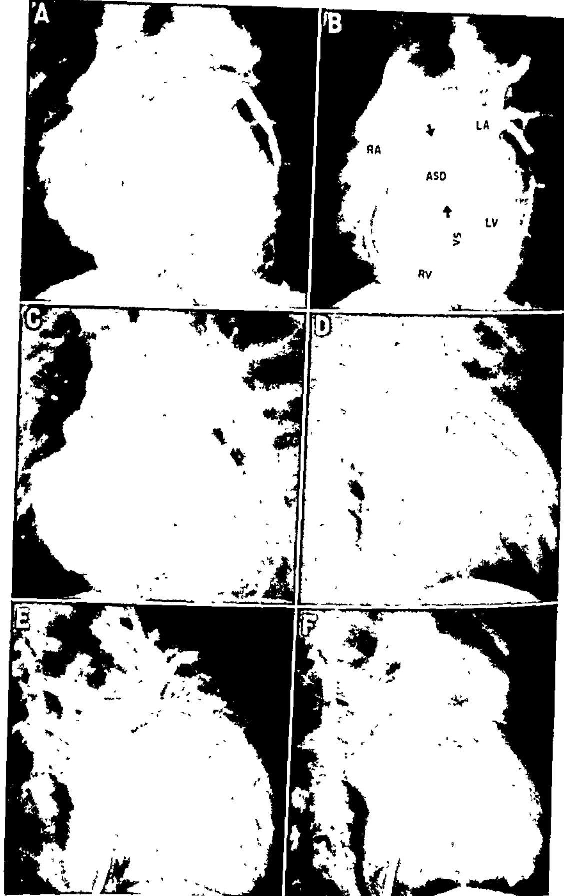


Fig. 419 (legend on facing page)

any event, no definite views can be expressed with regard to the size of the defect or its exact position This holds good even when the injection is made into the right ventricle or the pulmonary artery, as was fully evident in six of our cases. Furthermore, even if the contrast medium can definitely be shown to appear in the right atrium during recirculation, the possibility remains that it has been transported by pulmonary veins with aberrant openings.

Under these hemodynamic conditions, it seems that an atrial septal defect is visualized best if the contrast medium is deposited in the left atrium, in which case the opacified stream passing through the septum will indicate the approximate size and site of the defect.

We have found that even if contrast medium is injected rapidly into the atrium through a conventional catheter provided only with a hole in the tip, the results are unsatisfactory. Mixing with the blood in the left atrium is incomplete, and reflux to one or several of the pulmonary veins is common and is apt to decrease still further the desired local density beside the septum (Fig. 419).

One solution of the problem is to inject the contrast medium so that it is directed toward the septum By blocking the opening in the tip of the catheter and allowing the medium to run out through multiple holes in the sides instead, the greater part of it can be expected to follow the flow in the direction of the right atrium. In the Lehman type of catheter, all the lateral holes were bored perpendicular to the longitudinal axis of the catheter. Experience with this method has shown that the injection must be given rapidly, if the desired density is to be achieved, it should preferably be [illegible]

I [illegible]

duc [illegible]

through the atrial septal defect from below. As a rule the catheter then advances in the left atrium in a smooth curve, and the injection is made at a certain distance above the atrioventricular border. The movements of the catheter during injection are minimal, and the contrast medium is more easily taken up by the blood flow into the right atrium.

If the catheter is introduced through an arm vein, its tip is propelled toward the atrioventricular border during injection, owing to [illegible]

[illegible]

left ventricle, with the risk of the septal defect being incompletely outlined and the septum being [illegible]

it [illegible]

the examination. We have found that the best information is provided by exposures in the oblique projection, corresponding to an angle of 45°.

Owing to the short injection time, a rapid change of exposures must be made This requires a short exposure time. In our examinations, we have considered it desirable to use a frequency of 12 pairs of exposures per sec and an exposure time of 3/1,000 (1 330) sec In older children and adults the rate must, however, be reduced to six pairs of exposures per sec, because the roentgen tube does not tolerate the necessary increase in load per time unit Since, in the presence of an ostium primum defect in particular, it has proved necessary for exposures to be made in both ventricular systole and ventricular diastole, the lower frequency of exposures is not invariably adequate. This technique has been worked out successively and has been applied in a total of 55 cases (639).

In order to evaluate the site and size of the defect, it is necessary to make a series

← Fig 419.—Same case as in Figure 418 [illegible] atrium A–C left [illegible] identify th[illegible] pulmonary [illegible] and B, 2/1[illegible] and RV, lef[illegible]

of exposures of the initial stage of the injection phase (Fig. 354, *A* and *B*). The serial changer must therefore be started before the injection is begun. The septal defect can generally be studied only on pictures obtained before opacification of the left ventricle has taken place. This is because, in the axial projection, its infundibulum and the aortic root are directly superimposed on the atrial septum, so that it is practically impossible to make any evaluation. When the atrial septal defect is large, the greater part of the contrast medium is usually borne into the right atrium by the blood stream, only a small proportion passing into the left ventricle. The smaller the septal defect, the smaller is the quantity of contrast medium carried into the right atrium and the greater is the opacification of the left ventricle. When the foramen ovale is patent, there is either no leakage to the right atrium (Fig. 420) or only inappreciable leakage (Fig 445b).

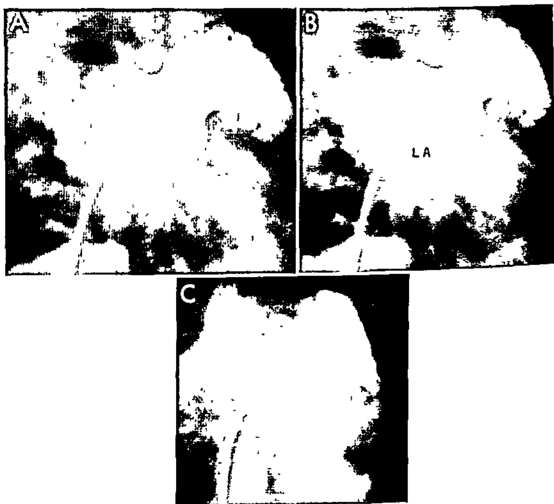


Fig. 420.—Patent foramen ovale and ventricular septal defect. Boy, aged 1 (M Z. 541103) Injection into left atrium (*LA*) shows no passage of contrast medium to right atrium Atrial septum well delimited (*C*) Position and course of the catheter typical of those in a defect at the site of the foramen ovale *A* and *B*, right anterior oblique projection *C*, left anterior oblique projection.

Figures 421–436 show part of the findings in some cases in which the method was successful, owing to use of a specially designed catheter and a rapid series of exposures. The pictures show that the contrast-mixed blood passes through the septum, as a stream of varying width, into the right atrium, mixing with the blood flowing in from the venae cavae and coronary sinus

The continued use of this angiocardiographic technique has shown that it contributes to more exact evaluation of the nature of the defect, provided that injection of the contrast medium is sufficiently rapid and that satisfactory opacification is obtained Forty-five of the 55 cases examined fulfilled these requirements. The remaining 10 could not be judged in this respect, but the examination confirmed the presence of an atrial septal defect. The essential causes tainty in these cases as in those in which operation was done under direct vision in the open heart.

The angiocardiographic observations described hereafter refer to cases in which operation was performed or the diagnosis verified at autopsy.

A *foramen ovale defect* was considered, on the basis of the operative findings, to be present in 20 cases On the angiocardiograms, the defect was situated in the pos-

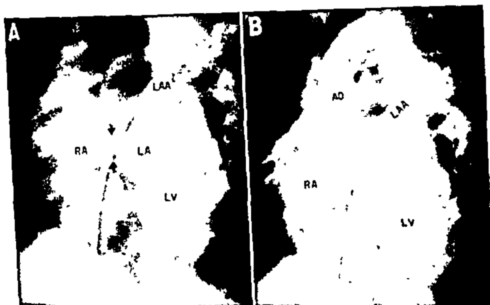


Fig 421.—Atrial septal defect of foramen ovale type Girl, aged 5 (M A 481009) The defect, which is fairly small, lies between the arrows in *A* The greater part of the contrast medium passes into the left ventricle and only a small quantity into the right atrium No operation performed *AO*, aorta *LA* and *RA*, left and right atria, *LAA*, left auricular appendage, *LV*, left ventricle

of an unsatisfactory quality of the angiocardiograms were unduly slow injection of contrast medium, because a sufficiently wide-bore catheter could not be introduced, and recoil of a catheter, provided only with a hole in the tip, into the right atrium

In 32 of the aforementioned 45 cases, the angiocardiographic findings could be compared with those at operation The greater proportion of atrial septal defects were, however, repaired by circumclusion, the blind technique being used Consequently, evaluation of the anatomic details presumably cannot have the same degree of cer- terior part of the atrial septum, usually just above the upper border of the inferior vena cava (Figs 423–426) For, the catheter inserted through the defect generally advanced into the left atrium in the direction of the left superior pulmonary vein This finding nevertheless applied only if the catheter was inserted through the defect and did not pass through a patent foramen ovale A more or less marked, thin ridge was present between the septal defect and the base of the ventricle (Figs. 425 and 427), representing the basal remains of the septum If the defect was so large that it

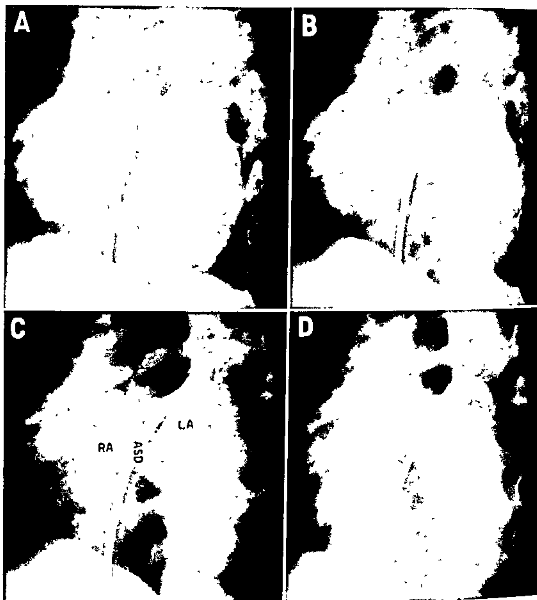


Fig. 422a.—Same case as in Figure 421, left anterior oblique position Illustrates the neces- [illegible] the atrial septal defect 1/12 [illegible] thin ¾ sec; 3/12 sec after be- [illegible] to visualize the defect *AO*, aorta, *ASD*, atrial septal defect, *LA* and *RA*, left and right atria; *LV* and *RV*, left and right ventricles, *VS*, ventricular septum (*continued*)









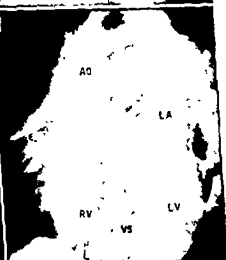


Fig. 422a (cont)

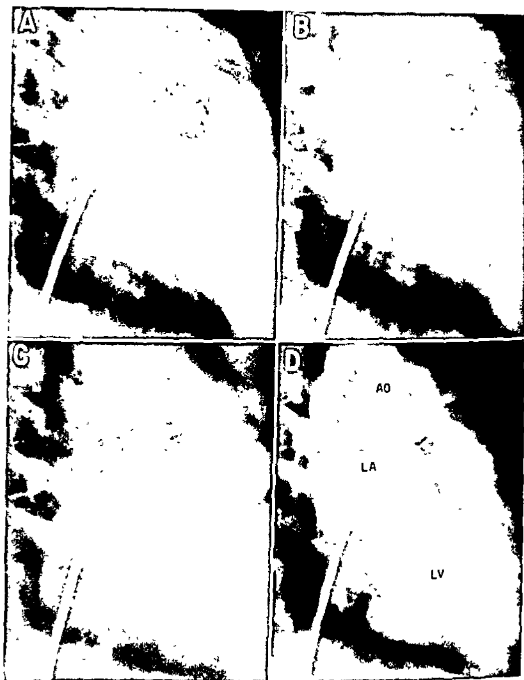


Fig. 422b.—Same case as in Figures 421 and 422a, right anterior oblique position *AO*, aort
LA, left atrium, *LV*, left ventricle

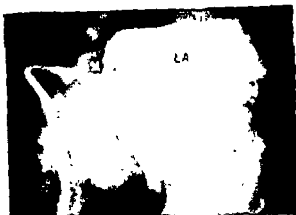


Fig 423.—Atrial septal defect of foramen ovale type Girl, aged 8 (B.A. 471001). Since the septum is not depicted altogether tangentially, the lower margin of the defect is not fully distinct Lack of interatrial passage of contrast medium at the base of the ventricle nevertheless argues against the presence of a septum primum defect *LA* and *RA*, left and right atria

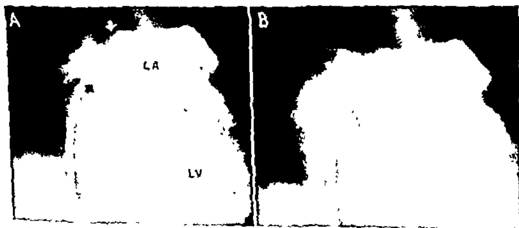


Fig 424.—Small atrial septal defect of foramen ovale type, Girl, aged 3 (I.N 531015). Upper arrow points to superior part of septum and lower arrow to lower margin of defect. *LA*, left atrium, *LV*, left ventricle

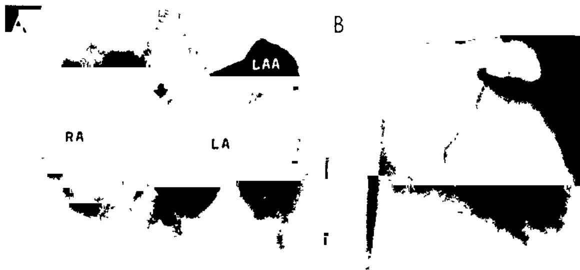


Fig. 425.—Atrial septal defect of foramen ovale type. Girl, aged 5 (A-B N. 510418). Large defect (between arrows) at site of foramen ovale. The catheter has advanced into the typical position in this type of defect: far to the rear and upward *LA*, left atrium, *LAA*, left auricular appendage, *RA*, right atrium.

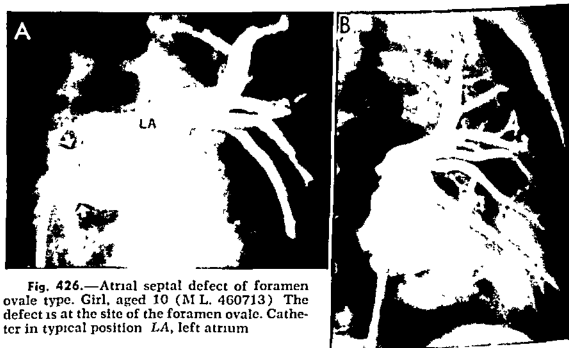


Fig. 426.—Atrial septal defect of foramen ovale type. Girl, aged 10 (M L. 460713) The defect is at the site of the foramen ovale. Catheter in typical position *LA*, left atrium

extended almost to the base of the ventricle, it was difficult to distinguish it from an ostium primum defect.

In one case, in which operation disclosed a defect of the *inferior vena cava type*, the most remarkable angiocardiographic feature was the rapid, intense opacification of this vein, indicating direct passage of blood from the left atrium to the inferior vena cava (Fig 428).

A *sinus venosus defect* was present in two cases On the angiocardiograms, it was depicted high up in the atrial septum, at the level of the orifice of the *superior vena cava* (Figs. 429 and 430). A *typical feature of* this kind of defect was that the stream of contrast medium followed the superior wall of the atrium into the right atrium, without *being interrupted* by any protruding remains of the septum This typical localization of the defect would seem *to* provide sufficient *grounds for the* aforementioned special diagnosis, even though the *aberrant* opening of the right pulmonary veins invariably present in this lesion cannot be demonstrated on the angiocardiograms

An *ostium primum defect* was present in 10 cases The most typical angiocardiographic feature was that the contrast medium, during its passage from left to right atrium, followed the base of the ventricle in an even stream, unbroken by any remains of the septum bulging into it (Figs. 431–434). As a result, its course was usually sharply delimited from the atrioventricular border by a fairly straight margin (Figs. 434 and 435). In one of the cases there was a coincident small defect in the uppermost part of the ventricular septum. This appeared on the angiocardiograms as a small hollow between the atrioventricular valves bulging toward the atria in *ventricular systole* (Fig 435). In three cases the possible existence of a low ridge in the basal *part of the* atrial septum could not be ruled out by angiocardiography. At operation, a thin ridge, which did not permit a suture to be anchored, was found in only one of these cases. The catheter advanced through the atrial septal *defect* generally passed through the left atrium far more basally than in cases with a foramen ovale defect. *The catheter tip* was usually observed caudad *to the left* auricular appendage.

Unusual features were observed in the case illustrated in Figure 436.

The size of the defect showed a wide range of variation in both the ...

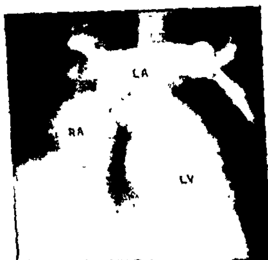


Fig 427.—Atrial septal defect of foramen ovale type Boy, aged 6 (C G. 510526). *The central rays are directed 30° caudally and the patient rotated 45°* Defect (between arrows) at site of foramen ovale extends almost to base of ventricle. *LA*, left atrium; *LV*, left ventricle, *RA*, right atrium

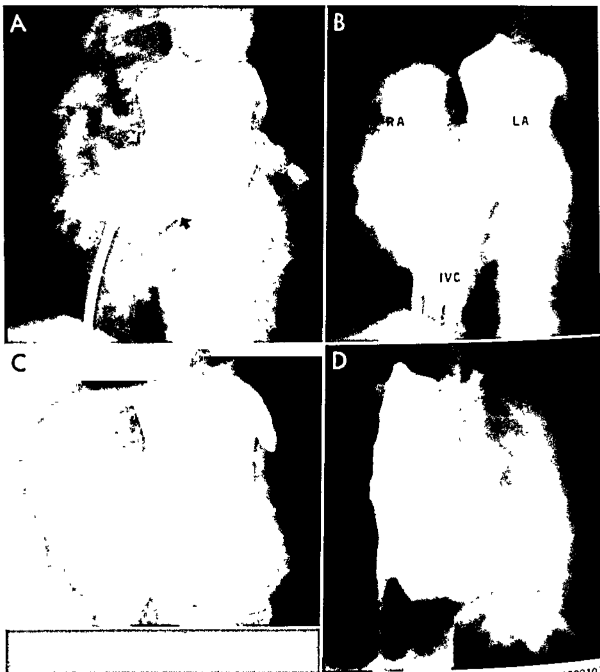


Fig. 428.—Large atrial septal defect of inferior vena cava type Boy, aged 16 (E B 400219) The septal defect (between arrows) extends far dorsally (see position of catheter in *D*) and into the inferior vena cava Part of the stream of contrast medium flows directly into this vessel (*A* and *B*). *IVC*, inferior vena cava, *LA* and *RA*, left and right atria

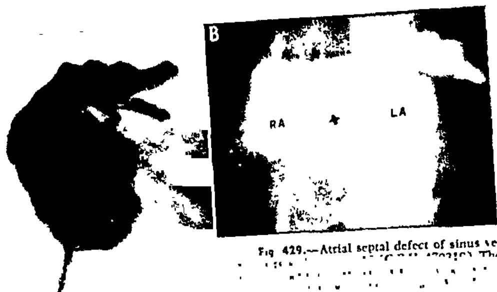


Fig 429.—Atrial septal defect of sinus ve- [illegible] The [illegible] left and right atria.

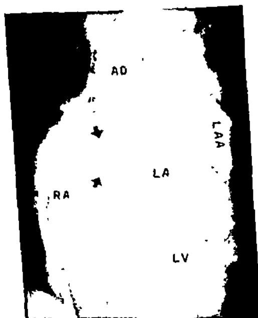


Fig 430 —Atrial septal defect of sinus venosus type Girl, aged 13 (B J 430726) The defect (between arrows) is directly below the orifice of the superior vena cava. *AO*, aorta, *LA*, left atrium, *LAA*, left auricular appendage, *LV*, left ventricle, *RA*, right atrium.

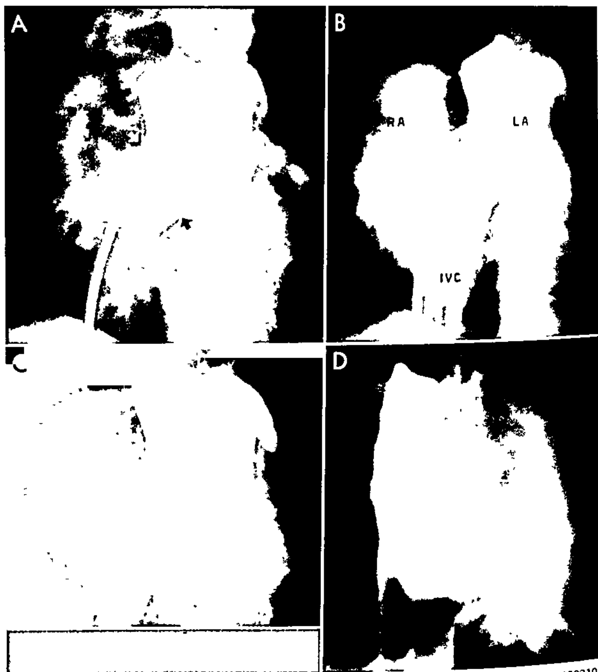


Fig. 428.—Large atrial septal defect of inferior vena cava type Boy, aged 16 (E B 400219) The septal defect (between arrows) extends far dorsally (see position of catheter in *D*) and into the inferior vena cava Part of the stream of contrast medium flows directly into this vessel (*A* and *B*) *IVC*, inferior vena cava, *LA* and *RA*, left and right atria

septum secundum and septum primum defects On technical grounds, the width of the defect could be estimated only in the left anterior oblique view. The dimensions given at operation were generally somewhat larger. We have been unable to establish the cause of this discrepancy. In several cases the basal ridge in the defect was poorly visualized, possibly because the atrial septum was not depicted tangentially enough. It must also be borne in mind that the basal ridge, seen in the left anterior oblique view, is somewhat distorted on deof the atrium. The course of the catheter sometimes facilitates the interpretation, by suggesting a possible anterior or posterior margin of the defect. There is, however, reason to assume that in several of our cases the catheter did not pass along the margin of the defect, but more in the center of it. Moreover, the extent to which the catheter can deform the margins of the septum is unknown. In none of our cases did multiple defects appear on the angiocardiograms.

It must also be mentioned that regurgita-

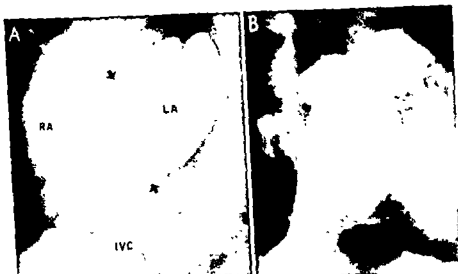


Fig 432 —Large atrial septal defect of ostium primum type Girl, aged 8 (E-M M 461105) The defect (between arrows) extends to the base of the ventricle The catheter runs just above the atrioventricular border (*B*) *LA* and *RA*, left and right atria, IVC, inferior vena cava.

piction, owing to the slope of the atrioventricular plane and that it includes those parts of the basal septum not involved by the defect. In some cases we tested exposures in the left anterior oblique view with the rays directed about 30° caudally, in order to depict the ridge tangentially to the atrioventricular plane (Figs 427 and 436) The value of this depiction is, however, limited both by superimposition of the heart on the liver and by the unavoidable distortion in depiction in the other plane

In the right anterior oblique view, the septal defect cannot be expected to stand out sufficiently sharply from the other parts tion to pulmonary veins, resulting in their opacification, did not occur unless part of the catheter lay in the orifice of any of the veins or in its immediate vicinity Consequently, pulmonary veins with anomalous drainage into the right atrium were not visualized in these examinations

On films exposed in the right anterior oblique view, an approximately tangential depiction of the mitral orifice is obtained. In every case we considered that we were able, on the basis of the angiocardiographic observations, to rule out associated mitral stenosis In one of our cases in which mitral stenosis was found at operation ...

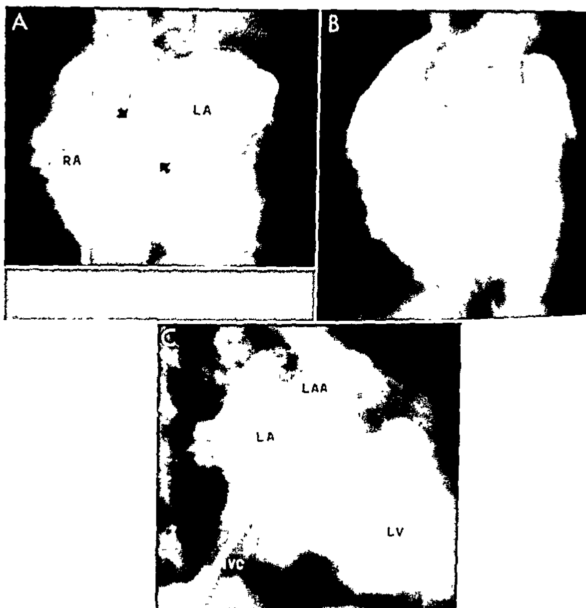


Fig. 431.—Atrial septal defect of ostium primum type. Girl, aged 8 (M.O. 470615). The defect (between arrows) is situated far down and anteriorly in the septum. The catheter, which impinges on the upper margin of the defect (*A*), lies in a plane anterior to the left auricular appendage, slightly above the atrioventricular border (*C*). *IVC*, inferior vena cava, *LA* and *RA*, left and right atria, *LAA*, left auricular appendage, *LV*, left ventricle.



Fig 434 —Atrial septal defect of ostium primum type. Boy, aged 11 (K-O K 431119) The large defect extends to the base of the ventricle No remains of the septum are visible basally. LA and RA, left and right atria.

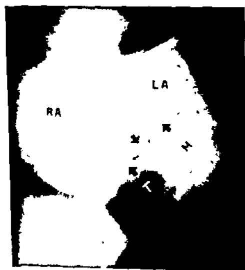


Fig 435.—Atrial septal defect of ost[illegible]

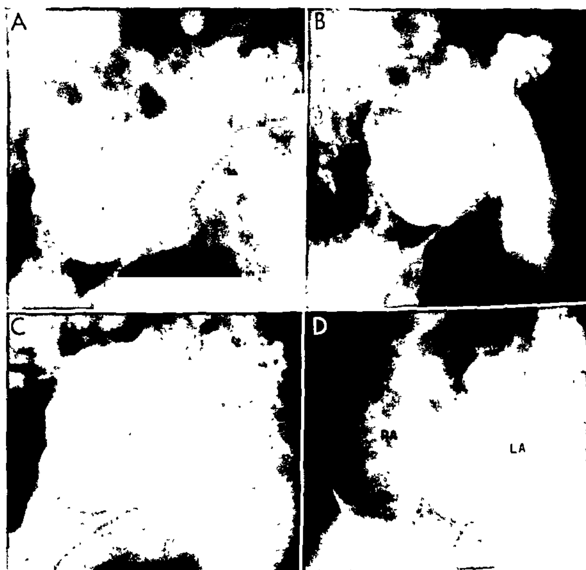


Fig. 433.—Large atrial septal defect of ostium primum type Woman, aged 34 (A N 211118) The catheter follows the atrioventricular border (*A–C*) The defect lies directly above the ventricular septum Its size is difficult to evaluate (*D*) owing to insufficiently oblique projection *LA* and *RA*, left and right atria.

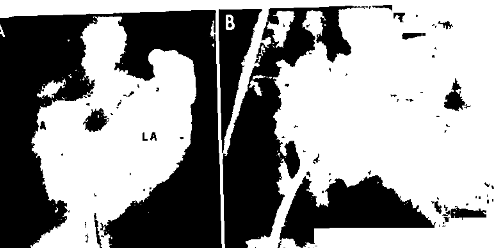


Fig 434.—Atrial septal defect of ostium primum type. Boy, aged 11 (K-O K 431119). The large defect extends to the base of the ventricle. No remains of the septum are visible basally. LA and RA, left and right atria



Fig 435 —Atrial septal defect of ost... fect lies far basally, its upper ... which the upper part is sligh... into a small hollow (arrow) ... which bulge upward in ventr... systole LA and RA, left and right atria

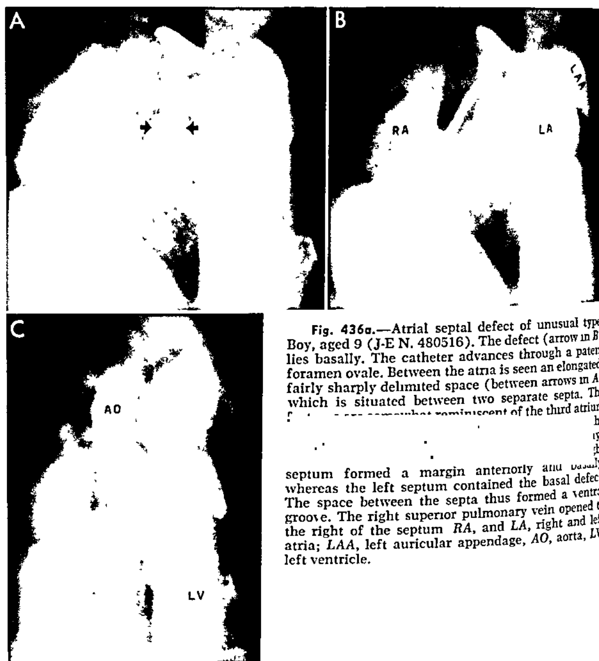
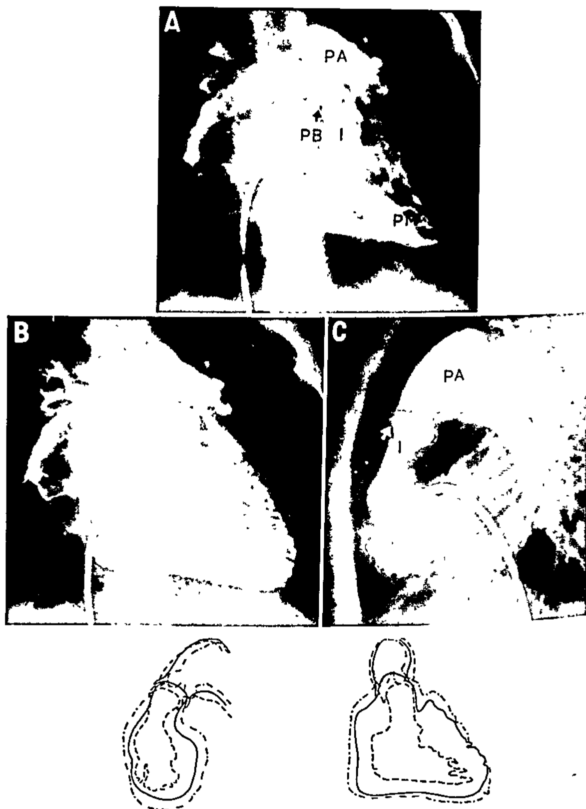


Fig. 436a.—Atrial septal defect of unusual type Boy, aged 9 (J-E N. 480516). The defect (arrow in *B*) lies basally. The catheter advances through a patent foramen ovale. Between the atria is seen an elongated, fairly sharply delimited space (between arrows in *A*) which is situated between two separate septa. The [illegible] somewhat reminiscent of the third atrium [illegible] he [illegible] ys [illegible] ght septum formed a margin anteriorly and basally, whereas the left septum contained the basal defect The space between the septa thus formed a ventral groove. The right superior pulmonary vein opened to the right of the septum *RA*, and *LA*, right and left atria; *LAA*, left auricular appendage, *AO*, aorta, *LV*, left ventricle.



Fig 436b —Same case as in Figure 436a Contrast medium injected into the left ventricle to rule out a complicating septal defect below the valves No regurgitation of contrast medium to left atrium *AO* ascending aorta *I*, infundibulum, *LV*, left ventricle



Late systole - - - -   Late diastole — — —   Early systole ———

Fig. 437.—Atrial septal defect and valvular pulmonary stenosis Boy, aged 4 (L-O B [illegible] (arrows) with a central orifice The *CSV*, crista supraventricularis, *I*, [illegible], papillary muscle, *SB*, septal band



Fig. 438 —Atrial septal defect and valvular pulmonary stenosis Man, aged 48 (Y.C. 050912) Apart from hypertrophy of the right ventricle (*B*), the conventional roentgenogram does not differ from that in uncomplicated atrial septal defect (*A*) Cusps of the pulmonary valve are fused into a dome (arrows in *B*) with a central orifice *CSV*, crista supraventricularis, *I*, infundibulum, *PA*, pulmonary artery

diographic examination had not been made

Incompetence of the atrioventricular valves cannot be established by this method, but requires separate injection of contrast medium into the ventricles if it is to be demonstrated on angiocardiography In such examinations, lack of regurgitation of contrast medium into the atrium presumably rules out incompetence of the valves (Fig 436b) Positive findings must be interpreted with caution, in view of the fact that closure of the valves may be prevented by the catheter Grant *et al* (298) have chosen to introduce the catheter into the left ventricle via the aorta, which would seem to obviate this source of error.

In four of our cases, associated valvular pulmonary stenosis was visualized by injection of contrast medium into the right ventricle. The stenosis presented the typical appearance of a dome with a central orifice (Figs 437 and 438). In two other cases, in which angiocardiographic examination was performed by injection into the left atrium, the contrast medium was so greatly diluted during passage into the right atrium and ventricle that the stenosis could not be identified Slight infundibular stenosis was visualized in one case

13 Persistent Ostium Atrioventriculare Commune

PERSISTENT OSTIUM atrioventriculare commune is a malformation arising through underdevelopment of the dorsal and ventral atrioventricular endocardial cushions, so that they fail to fuse. As a result, the embryonic atrioventricular canal persists, and fusion of the septum primum with the atrioventricular cushions does not take place. The omission of the latter process causes a large defect in the lower part of the atrial septum, corresponding to the ostium primum, as well as in the superior region of the ventricular septum, in its membranous part The atrial septal defect has a typical crescentic shape, and the septum stretches in an arch across the atrioventricular canal The cusps are invariably deformed Both the anterior mitral cusp and the septal tricuspid cusp are cleft as far as the base This cleft continues through the superior part of the ventricular septum. Sometimes there is a common atrioventricular valve, with an anterior and a posterior cusp, which hang down into both ventricles (Fig. 443b). The differentiation from ostium primum defects has been discussed on page 415.

The atrioventricular cusps are usually incompetent Consequently, there is not only a shunt between the right and the left side of the heart, but also regurgitation from the ventricles to the atria. Since the chambers of the right side of the heart offer lower resistance to filling than those of the left side, a dominantly left to right shunt takes place. Presumably, the blood is shunted not merely from the left atrium to the right and from the left ventricle to the right, but also directly from the left atrium to the right ventricle during ventricular diastole, and from the left ventricle to the right atrium during ventricular systole. If the defect is extremely large, there are possibilities of a bidirectional shunt, and in cases with high pulmonary vascular resistance a dominantly right to left shunt can occur.

This malformation is severe and often leads to heart failure at an early age. More than half the patients die during their first year of life and only a few survive until adult age (216, 138, 502, 680). The malformation is frequently associated with mongolism (216, 265, 649).

We have seven such cases in our series. mongolism was present in one of them. Autopsy was performed in four cases Three patients are still alive.

CLINICAL FEATURES

The chief symptoms and physical findings in our cases may be inferred from Table 9. The course was severe, with early appearance of cardiac failure and death in four cases. All the patients were incapacitated and physical development was usually retarded Cyanosis was present in all but one case, but was not prominent. All had

TABLE 9.—PERSISTENT OSTIUM ATRIOVENTRICULARE COMMUNE SYMPTOMS AND PHYSICAL FINDINGS IN 7 CASES

CASE	AGE AT DEATH	HISTORY AND GENERAL CONDITION	CYANOSIS	DYSPNEA	CARDIAC FINDINGS
M E 500121 (girl)	4 yr, 4 mo	Heart disease detected at age 4 mo; development normal, since age 4, unable to play like other children, past 2 months, successive deterioration, confined to bed, edema and liver enlargement	Slight in recent months	Mod	Precordial bulge; parasternal lift, acc. 1st sound; acc. 2nd sound over pulm area, systolic murmur (grade 4), max over apex, peripheral pulses weak
B.N 530908 (girl)	1 yr	Poor weight gain, cannot stand with support but can sit, 1 attack of unconsciousness at 6 mo, edema earlier, disappeared after digitalization	Slight since age 2 mo	Mod	No precordial bulge; acc. 2nd sound over pulm area; no murmur; peripheral pulses weak
G B 540421 (boy)	2 mo	Mongoloid, hospitalized from birth until death, poor weight gain, gradual deterioration, last few weeks, too feeble to take the breast, tube fed	Slight, disappeared on breathing O_2	Mod.	From age 2 wk, moderately loud systolic murmur to right of sternum, peripheral pulses strikingly weak
L O F. 461018 (boy)	Alive 8 yr, 7 mo	Retarded physical development, fatigued during slight exercise, unable to play like other children, never edema	Slight, periodic	Mod. on exertion	Precordial bulge; parasternal lift; thrill, acc 2nd sound and grade 3 systolic murmur over pulm area; pansystolic murmur over apex transmitted to left axilla; mid-diastolic murmur over apex
I K. 530414 (girl)	Alive 2 yr, 10 mo	Poor weight gain first year, since then improvement, started to walk at 18 mo, only slightly incapacitated, no edema, but liver enlargement	0	Slight on exertion	Precordial bulge, heaving apex beat, acc 2nd sound over pulm area, pansystolic and mid diastolic murmur over apex
B D 560308 (boy)	7 mo	Poor weight gain, acute heart failure at age 7 mo, with pulmonary edema and liver enlargement	Slight at age 7 mo	Mod since age 3 mo	No precordial bulge; grade 2 systolic murmur over apex, peripheral pulses weak
A.K. 520426 (girl)	Alive 3 yr, 1 mo	Repeated respiratory infections, unable to run, no edema, but liver enlargement	Slight since birth	Mod. on exertion	Slight precordial bulge; parasternal lift; acc. 2nd sound and systolic murmur over pulm area, pansystolic and mid-diastolic murmur over apex

dyspnea, either at rest or on exertion The second sound over the pulmonary area was accentuated In one case there was no murmur, but in most of the others a systolic murmur was heard over the pulmonary area, indicating an increased pulmonary flow, as well as an apical systolic murmur, caused by regurgitation in the atrioventricular valves The peripheral artery pulse was small in the most severe cases, as an expression of a low output in the systemic circulation

ELECTROCARDIOGRAPHY.—In ostium primum defects, both with and without interventricular communication, the electrocardiogram is often characteristic, with an incomplete right bundle-branch block in combination with left axis deviation (cf. p

424). In persistent ostium atrioventriculare commune, the interventricular communication usually results in high pressure in the right ventricle, consequently, electrocardiographic signs of right ventricular hypertrophy are often found (Fig. 439). In cases with mitral regurgitation, left ventricular hypertrophy may also be present.

In our series left axis deviation was present in five cases. Thus this feature was lacking in two cases, in which the diagnosis was verified at autopsy. An incomplete right bundle-branch block was present in five cases, hypertrophy of only the right ventricle in three, hypertrophy of only the left ventricle in one, and combined hypertrophy in two cases. High, peaked P waves were recorded in four cases. A prolonged P-R interval has often been found (656), but in our series the interval was normal.

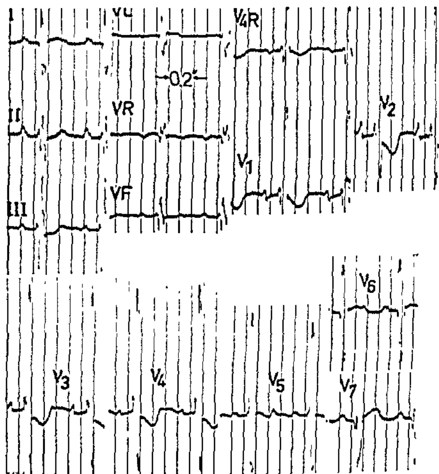


Fig. 439.—Electrocardiogram in persistent ostium atrioventriculare commune Girl, aged 4 (M.B. 500121). Cardiac failure was present, and she was taking digitalis.

ROENTGENOLOGIC EXAMINATION

The roentgenologic findings in this cardiac malformation have only rarely been commented on in the literature (24, 488, 566, 650).

The roentgenologic appearance in four of our seven cases is illustrated in Figures 440–442. It was characterized by enlargement of the right side of the heart, dilatation of the main trunk of the pulmonary artery and its central and peripheral branches, and a considerably increased heart volume. In contrast to other writers (265), we do not consider that any special features are present in the frontal view which permit a differential diagnosis from uncomplicated atrial septal defect.

The dilatation of the right atrium was marked and was most pronounced in the



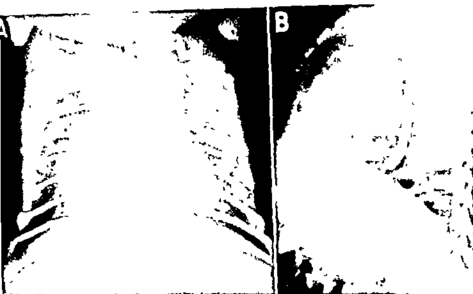




Fig. 440a —Persistent ostium atrioventriculare commune Girl, aged 4 (M.B 500121). Heart volume increased to more than double, great enlargement of right atrium and ventricle, long contiguity of surface of ventricle to anterior wall of thorax, greatly increased vascularity of lungs, no enlargement of left atrium



Fig. 440b.—Same case as in Figure 440a Large defect in caudal part of atrial septum (OAC in A and B) and in cranial parts of ventricular septum. Cusps of the mitral valve extend through the ventricular septal defect (the arrow in C) into the right ventricle and are partly fused with cusps of the tricuspid valve Medial and anterior cusps of the tricuspid valve are greatly malformed (arrow in A)—small, thick, nodulated, with short chordae tendineae. Right atrium is extremely large, and trabeculae greatly hypertrophied Wall musculature is also thickened Left atrium of normal size, no thickening of its wall Papillary muscles of the greatly enlarged right ventricle are fused and run in an abnormal direction, considerable hypertrophy of trabeculae and wall muscles Slight hypertrophy of trabeculae of left ventricle Cusps of mitral valve slightly thickened *LA* and *RA*, left and right atria, *LAA*, left auricular appendage, *LV* and *RV*, left and right ventricles, *MV*, mitral valve, *OAC*, persistent ostium atrioventriculare commune, *PM*, papillary muscle, *TV*, tricuspid valve, *VS*, ventricular septum



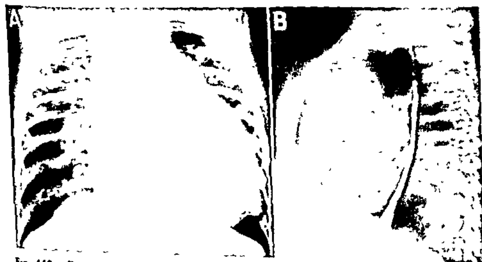


Fig. 442 —P...
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cases in which severe heart failure had developed.

There was increased contiguity of the right ventricle to the anterior wall of the thorax, as in dilatation. The apex was upturned in the cases in which enlargement and hypertrophy of both the right and the left ventricle were found at autopsy. The size of the left ventricle could not be estimated by roentgenologic examination in any of the cases. The main trunk of the pulmonary artery exhibited increased pulsations in those cases in which evaluation was not hampered by the thymus. Since most of the patients were infants, the width of the aorta was somewhat difficult to judge. Only in one case could the aorta be visualized clearly and was seen to be of ordinary width. In the other cases it was presumably narrow, this was confirmed in the four autopsied cases.

In four cases the left atrium was not enlarged, in two there was suggested enlargement, and in the remaining case the atrium was definitely dilated. Enlargement or lack of enlargement of the left atrium showed no definite relation to the presence or absence of mitral incompetence, studied by selective angiocardiography. Friedman and Ash (265), like Taussig (650), have pointed out that it is not uncommon to find no enlargement of the heart in asymptomatic patients. In most cases, however, the *enlargement is moderate or considerable.*

CARDIAC CATHETERIZATION

Since the communication between the right and left side of the heart is on the level of the atrioventricular plane, it may be of diagnostic value to make a careful study under fluoroscopy of the position of the catheter on passage through the defect (cf. p. 447). A patent foramen ovale is not infrequently present as well, and if the catheter passes through it, the malformation may be misinterpreted as a foramen ovale defect. A left to right shunt not only at atrial level, but also at ventricular level, has been stated to indicate a persistent ostium atrioventriculare commune (681). In our experience, however, this is often found in isolated atrial septal defect as well In our foramen ovale defects, an average 25 per cent of the left to right shunt took place at ventricular level. This is because, during the phase of rapid filling, the blood from the left atrium flows through the defect down toward the tricuspid orifice, without any effective mixing with the blood in the right atrium.

In persistent ostium atrioventriculare commune, pulmonary hypertension usually is present, and the arterial oxygen saturation is often decreased. Both these findings are unusual in uncomplicated atrial septal defect in children. Consequently, if a left to right shunt, mainly at the atrial level, is found in combination with pulmonary hypertension and somewhat decreased arterial oxygen saturation, the first suspicion should be of a persistent ostium atrioventriculare commune. Selective angiocardiography is, however, necessary for a definite anatomic diagnosis.

Cardiac catheterization was performed in six of our cases. Table 10 shows the results in five of them. In case M.B. 500121, there was severe cardiac failure, the pressure was raised in both the right and the left atrium (13 and 16 mm *Hg*, respectively), as was the pressure in the right ventricle, which was systemic. The *blood-gas* analysis did not provide any *reliable* data regarding the shunts, owing to the poor condition of the patient, with irregular respiration and periodic cardiac arrhythmia The arterial oxygen saturation was exceedingly low (52 per cent).

The right ventricular pressure was raised in every case. In two cases *the catheter* could not be advanced into the pulmonary artery, but the presence of pulmonary stenosis could nevertheless be ruled *out* This was partly because the *PCA pressure* was high and partly on account of the physical signs and the angiocardiographic *findings*

The left to right shunt was large, and in all but one case a moderate *right to* left shunt was also present.

TABLE 10.—Persistent Ostium Atrioventriculare Commune: Findings on Cardiac Catheterization in 5 Cases*

Case		O_2 Content vol %								Arterial O_2 Sat, %	O_2 Cap vol %	Pressure, mm Hg								
													RV		PA				LV	
		SVC	IVC	RA	RV	PA	PV	LA	LV			RA Mean	Syst	End-diast	Syst	Diast	PCV Mean	LA Mean	Syst	End-diast
B N	530908	8 5	10 3	10 5	11 8	12 2	16 1	13 8	—	75	16 4	—	86	4	67	37	—	—	—	—
L O F	461018	10 4	—	12 2	—	—	16 0	15 2	14 5	89	16 7	5	—	—	—	—	—	4	96	5
	†	10 4	11 5	14 0	15 5	—	—	—	—	—	18 9	6	87	10	—	—	—	6	—	—
I K	530414	7 6	7 4	10 3	10 6	11 1	—	—	—	81	13 7	10	56	3	57	23	10	—	—	—
B D	560308	7 4	9 9	8 8	12 0	11 7	—	—	13 3	95	13 5	3	56	4	46	12	—	—	68	3
A K	520426	8 5	10 4	11 3	12 3	—	14 8	12 3	12 8	82	15 8	5	64	10	—	—	—	9	69	9

*For abbreviations see Table 1, p 119 †Data from catheterization performed one year earlier

ANGIOCARDIOGRAPHY

At venous (peripheral) angiocardiography, rapid, lengthy opacification of all the chambers of the heart is a characteristic feature, as pointed out by Gasul (272), Goodwin *et al.* (286), and Abrams and Kaplan (4). It is obvious that no details of the pathologic anatomy beyond those apparent from ordinary roentgenologic examination are disclosed by this procedure.

A thorough, selective examination implies that both the atrial and the septal defect are depicted. Experience shows that this requires two injections of contrast medium, one into the left atrium and the other into one of the ventricles. We performed selective angiocardiography in six of our cases, but confined the examination to a study of the ventricle, since the existence of an atrial septal defect had been fully evidenced at catheterization. The catheter for injection was introduced into the left ventricle in three cases and into the right ventricle in the other three. In two of the cases, the movement of the catheter during injection permitted the direct conclusion that the atrioventricular orifice was a common one. For, in one of them the catheter tip passed from the right ventricle to the left during angiocardiography (Fig. 443a), and in the other it passed in the opposite direction.

In two of the three cases in which left ventricular injection was made, the ventricular septal defect was directly visualized (Fig 444b), and in one of them definite reflux to the left atrium was established. In the third case, the catheter was rapidly repelled into the right atrium at the beginning of injection, so that all four chambers of the heart were filled, and then appeared during prolonged opacification.

When the contrast medium was injected into the *right* ventricle, the ventricular septal defect was clearly visualized in two cases (Fig 443a), and a distinct reflux to the left atrium was observed in one of them Severe cardiac failure was present in the remaining case Emptying of the greatly

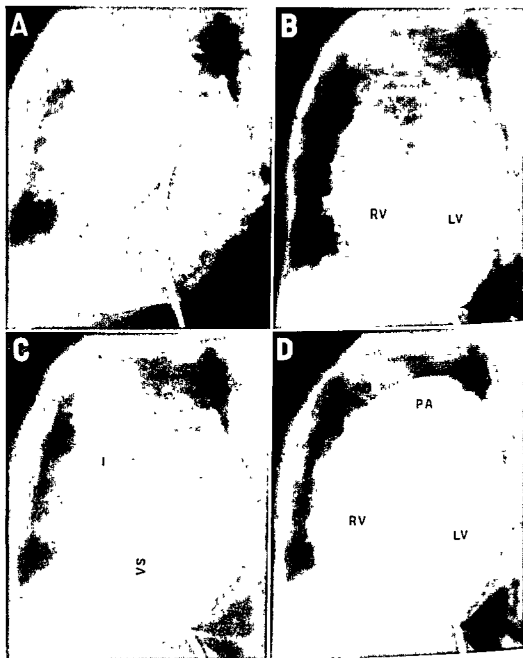


Fig. 443a.—Persistent ostium atrioventriculare commune Girl, aged 1 year (B N. 530908) *A* and *E*, the catheter, introduced into the right atrium, advances into the left ventricle and continues, via the ventricular septal defect, into the right ventricle. *B* and *F*, during injection, the catheter tip recoils into the left ventricle Most of the contrast medium in both ventricles passes into the pulmonary circulation (*C* and *G*, *D* and *H*), only a small quantity passes into the narrow ventricles are large, but the right is larger than the left left and right ventricles, *PA*, pulmonary artery, *VS*, ven-

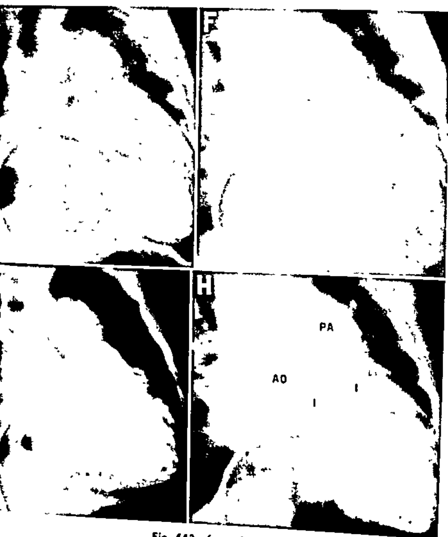


Fig 443a (*cont*)



Fig. 443b.—Same case as in Figure 443a. Large defect in caudal part of the atrial septum
(*ASD* ... communicate
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cusps are presen ... chordae tendineae ... he
cusps are attach ... re of the
same size, and ... powerful
... the left is of normal size. The pulmonary artery is wide and aorta (arrow in *D*)
... nd *SVC*, inferior and supe-
... l right ventricles, *PA*, pul-
monary ... valves, *VSD*, ventricular
septal defect.

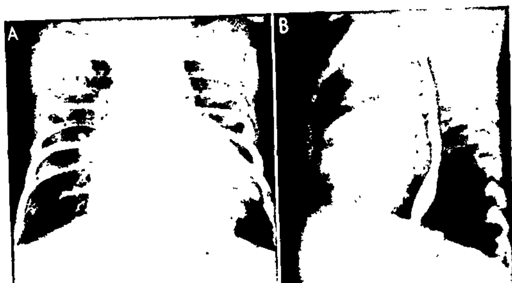


Fig 444a —Persistent ostium atrioventriculare commune. Girl, aged 6 months (B P. 570331). Considerable enlargement of the heart with great dilatation of right ventricle and slight enlargement of left Right ventricle has hypertrophic shape. Greatly increased blood volume in pulmonary circulation



Fig. 443b —Same case as in Figure 443a Large defect in caudal part of the atrial septum (*ASD*) and small defect at site of the foramen ovale (arrows in *A* and *B*) Atria communicate with ventricles through a common orifice Around the orifice are four cusps, two anterior and two posterior, their medial segments are partially fused, giving the impression that only two large cusps are present, one anterior and one posterior A large number of the chordae tendineae of the cusps are attached to the margin of a large ventricular septal defect. Both ventricles are of the same size, and thickness of their walls is the same The right atrium is large, with powerful trabeculation; the left is of normal size The pul[illegible] is wide and aorta (arrow in *D*) [illegible] *SVC*, inferior and supe[illegible] ight ventricles, *PA*, pul[illegible] alves, *VSD*, ventricular septal defect.

14 Communication Between Left Atrium and Coronary Sinus

SINCE the anterior cardinal vein is in intimate contact with the dorsal wall of the left atrium, a communication between this vessel and the left atrium can arise during *fetal life* It has not yet been elucidated whether this communication arises on account of abnormal development of the atrial sinus region, *or whether it* is due to an impediment to emptying of the cardinal vein in question, with resulting dilatation of a thebesian vein. In most of the cases described in the literature, there has been either a demonstrable *impediment to outflow* in the form of partial obliteration of the coronary sinus (43, 483, 520, 647), usually close to its entry into the right atrium, or constriction or *atresia of the left* innominate vein (43, 353). In the former case, the cardinal vein has opened into the left atrium through an orifice usually devoid of a valve (43, 353, 358). *A similar* anomalous entry of the left posterior cardinal vein has also been described (119) In cases in which pressure measurements have been made, higher pressure has been recorded *in the* veins drained by the left superior vena cava than in the rest of the venous system (353) This is because the pressure is higher in the left atrium than in the right

In those cases in which the coronary sinus has a free outflow to the right atrium, the anterior cardinal vein communicates with the left atrium, but drainage to the right atrium takes place via the coronary sinus. Consequently, there is no rise in pressure in the cardinal vein. Owing to the higher pressure in the left atrium, there is a left to right shunt to the coronary sinus The hemodynamic conditions are the same as in atrial septal defect. T ... nation ... ibed a ... addi-tional case in a thorough study of the literature. Weidman *et al* (691) mentioned the existence of this anomaly, but gave no references and reported no case of their own.

In one of our cases with mitral atresia and intact atrial septum, the left atrium was drained entirely through such a communication with the coronary sinus (see p 739) Our series also contains two cases in which the diagnosis could be made by means of cardiac catheterization and angiocardiography. One of the patients also had a functionally unimportant coarctation of the aorta, and in the other a persisting left superior vena cava opening into the coronary *sinus* was demonstrated. The salient features were as follows

BOY, AGED 10 YEARS (P H 450612) —Heart disease ...
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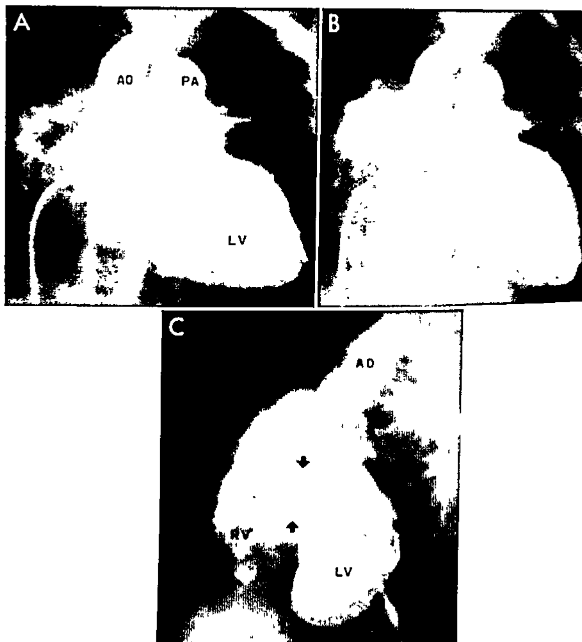


Fig. 444b.—Same case as in Figure 444a The catheter tip has passed from the right atrium into the left ventricle (*LV*) through an ostium primum and an atrioventricular orifice common to the two ventricles Large ventricular septal defect (between arrows in *C*) and passage of contrast medium into right ventricle (*RV*) Pulmonary artery (*PA*) somewhat wider than aorta (*AO*). *VS*, ventricular septum

dilated right ventricle took place so slowly in this case, and the contrast medium was so greatly diluted, that no diagnostic data of value were obtained.

In all except the last-mentioned case, a left to right shunt was demonstrated at the atrial level after passage of the contrast medium through the pulmonary circulation, providing additional evidence of a concomitant atrial communication.

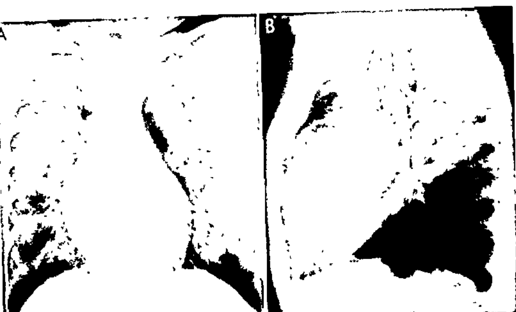


Fig 445a.—Communication between left atrium and coronary sinus. Boy, aged 10 (P.H 450612). Enlargement of right atrium and ventricle Moderately increased blood volume of lungs, with no marked enlargement of left atrium

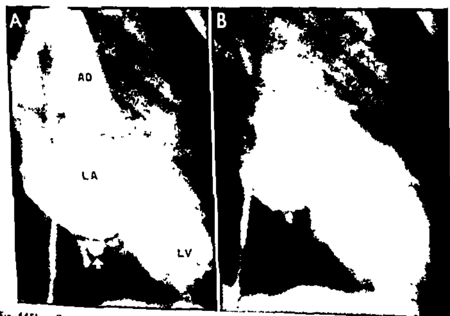


Fig 445b —Same case as in Figure 445a Catheter tip ad[illegible]
a patent f[illegible]

distinctly palpable. The electrocardiogram was normal.

GIRL, AGED 15 (E.J. 420909).—Heart disease was detected at a mass miniature-roentgenologic examination at age 12 years. She has never had any cardiac symptoms. Physical examination showed slight parasternal pulsations, a widely split second sound (0.08 sec), a grade 3, fairly high-frequency systolic murmur over the pulmonary area, and a low-frequency mesodiastolic murmur over the apex. The blood pressure was 125/80 mm Hg. The electrocardiogram showed an incomplete right bundle-branch block.

ROENTGENOLOGIC EXAMINATION

In both cases the roentgenologic appearance was the same as in uncomplicated atrial septal defect. It was characterized by enlargement of the right atrium and dilatation of the right ventricle, as well as by a dilated pulmonary artery with wide central and peripheral branches. A remarkable feature was that the left atrium was slightly dilated in one case. The presence of mild coarctation of the aorta in one case and a persistent left superior vena cava in the other could not be established until cardiac catheterization and angiocardiography were done.

CARDIAC CATHETERIZATION

In the first case (P.H.) the catheter passed into the left atrium through a patent foramen ovale According to angiocardiographic examination with injection of contrast medium into the left atrium, the valve covered the opening completely. Oxygen saturation of the blood in the left atrium and brachial artery was normal (96 per cent) A small left to right shunt was present at the atrial level. The oxygen saturation of the pulmonary artery blood was 78 per cent, and the ratio of pulmonary to systemic flow was 1 5.1 The coronary sinus was not catheterized, and the diagnosis was made on the basis of the angiocardiographic findings

In the second case (E J) the catheter could not be advanced from the right atrium into the left, although it had been introduced into a leg vein. From the left arm, the catheter passed through a persistent left superior vena cava into the coronary sinus and right atrium, and thence into the right superior vena cava. The oxygen saturation of the blood in both caval veins was the same (67 per cent). Far down in the left superior vena cava, at the opening of the coronary sinus, a great mixture of arterial blood was obtained (saturation 89 per cent). The catheter could be advanced from this position into the left atrium and thence into the left ventricle, where the blood had normal oxygen saturation (98 per cent). A large left to right shunt was present. The oxygen saturation of the blood in the right atrium, right ventricle, and pulmonary artery was 90 per cent, and the ratio of pulmonary to systemic flow was 2 8:1. The pulmonary artery pressure was normal (29/11 mm Hg). The diagnosis could be made on the basis of these observations, and the anatomic details of the malformation were demonstrated with the help of angiocardiographic examination.

ANGIOCARDIOGRAPHY

In one case (P.H.) the contrast medium was deposited in the left atrium. The injection was made into a catheter advanced through the foramen ovale. Leakage through the foramen ovale was inappreciable, and the absence of any further communication in the atrial septum was demonstrated. Early filling of the coronary sinus was observed, evidently via a communication with the left atrium. The actual communication was not visualized Opacification of the right atrium was due entirely to the inflow of contrast medium-mixed blood from the coronary sinus The passage of an interatrial shunt via the coronary sinus could thus be demonstrated by angiocardiography (Fig. 445).

In the other case (E.J.) the contrast medium was injected into the left superior vena cava, which emptied into the greatly dilated coronary sinus. Slightly proximal to the confluence of the flows in the sinus, great dilution of the contrast medium was

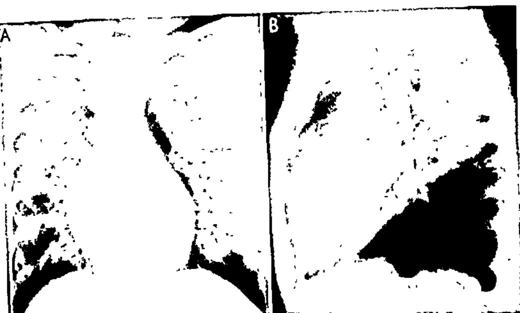


Fig 445a —Communication between left atrium and coronary sinus. Boy, aged 10 (P H 450612). Enlargement of right atrium and ventricle Moderately increased blood volume of lungs, with no marked enlargement of left atrium

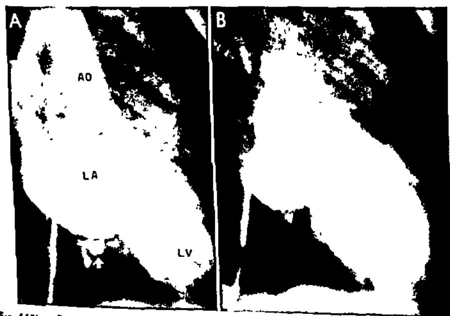


Fig 445b —Same case as in Figure 445a Catheter tip advanced into ...
a patent ...
nicatioı ...
LV, lef...



Fig. 446.—Communication between left atrium and coronary sinus Girl, aged 15 (E J 420909). Injection of contrast medium into left superior vena cava (*SVCs*). This vessel opens into a greatly dilated coronary sinus (*CS*), which communicates with the left atrium A large quantity of blood is shunted from the left to right atrium through this communication This causes the considerable dilution of contrast medium in the coronary sinus *RA*, right atrium

observed, it was due to inflow of blood unmixed with contrast medium from the left atrium (Fig. 446) After passage of the medium through the pulmonary circulation and opacification of the left atrium, the coronary sinus was once more visualized The left upper pulmonary vein opened to the right of the left superior vena cava; the other veins had the ordinary opening into the atrium These findings, together with the observations at cardiac catheterization, unquestionably indicated the presence of a communication between left atrium and coronary sinus.

15 Anomalous Drainage of Pulmonary Veins

IF ONE or several but not all of the pulmonary veins are drained into the right atrium or a systemic vein, the hemodynamic situation is identical with that in atrial septal defect. If, on the contrary, all blood from the lungs is drained into the right side of the heart, there is complete mixing of the arterial and venous blood. A communication between the right side and the left side of the heart then becomes a prerequisite for life

A. COMPLETE ANOMALOUS DRAINAGE

This malformation presents a characteristic syndrome, and detailed descriptions of its hemodynamics have been given in recent years (268, 382, 613, 697). The most common variant is drainage of the pulmonary veins into a persisting left superior vena cava, followed in order by drainage into the coronary sinus, the right atrium, the right superior vena cava, or, less frequently, the portal vein or the ductus venosus (99, 111, 178, 210, 215, 268, 382, 498, 596, 613, 697, 706). Such malformations are considerably more usual than was formerly supposed, formerly they were, in all probability, easily overlooked when autopsy was done.

Our series includes 11 cases In seven of them, the pulmonary veins were drained into a persisting left superior vena cava, which communicated with the right superior vena cava. In two cases, the pulmonary veins emptied into the coronary sinus, and in two cases, into the right atrium In addition, we had one case with complete anomalous venous return, mitral atresia, single ventricle, and transposition of the aorta, it is reported in detail in Chapter 25, on Mitral Atresia.

CLINICAL FEATURES

The malformation is severe and death often occurs before 1 year of age However, an increasing number of patients who have reached adult age have been reported on (111, 210, 268, 291, 489, 502, 596, 613, 643, 697) The prognosis is mainly determined by the size of the interatrial communication. The malformation can be compared to a common atrium in association with a varying degree of mitral stenosis, an intact atrial septum then corresponding to atresia of the mitral orifice and a very large atrial septal defect corresponding to a normal mitral valve.

The clinical features are also dependent on the size of the pulmonary flow. This is usually exceedingly large. The arterial oxygen saturation is then only inappreciably decreased and the patient is not cyanotic. The pulmonary flow is smaller in cases with complicating high pulmonary vascular resistance or pulmonary stenosis, and the

arterial oxygen saturation may be so low that cyanosis appears. Even these patients may reach a relatively high age, provided that the interatrial communication is so large that a relatively normal systemic flow can be maintained.

If the interatrial communication is very small, heart failure appears as early as the first year of life. Only a small proportion of the blood returned to the heart from the pulmonary and systemic circuits can pass through the atrial spetal defect. The stroke volume of the left ventricle is therefore were the main clinical features. Edema developed a few weeks before death. One patient was operated on at the age of 6 months, when heart failure had just started to develop. She died two days later. The autopsy findings in two of the cases may be inferred from Figures 451b and 453b.

The course was less severe in the other patients. One of them had been cyanotic since birth; in this case associated pulmonary stenosis was present. Cyanosis appeared in four cases after one year of age, but was only of mild degree. Two patients

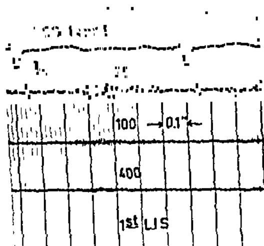


Fig 447.—Phonocardiogram in complete anomalous drainage of pulmonary veins Boy, aged 8 (M.M 460108) A typical continuous murmur is recorded over the first left interspace (L.I.S.) Boxed figures denote degree of amplification, other figures denote standard frequencies of the filters

small. Although the arterial oxygen saturation is high, the systemic flow is inadequate Right ventricular failure develops, the pulmonary flow then decreases, and arterial oxygen saturation falls The pressure rises in both the pulmonary and the systemic veins, and peripheral edema as well as pulmonary edema appears This state rapidly leads to death.

Four of our 11 patients had this severe course, with a fatal outcome during the first two years of life The heart disease was detected soon after birth, because of a murmur Poor weight gain, dyspnea, increased fatigability, slight cyanosis on exertion, and repeated [illegible]

had cyanosis on exertion only They all had dyspnea and increased fatigability of such degree that they were definitely disabled. No edema had been present.

The *cardiac findings* were characterized by a *precordial bulge*, parasternal lift, and a systolic murmur over the pulmonary area. The second sound over the pulmonary area was split, and the pulmonary component was accentuated In two cases a venous hum was also *audible*, it is a frequent occurrence *in this* malformation (382, 613). Figure 447 shows that the murmur is of the same nature as *that* in patent ductus arteriosus.

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If the interatrial communication is very small, heart failure appears as early as the first year of life Only a small proportion of the blood returned to the heart from the pulmonary and systemic circuits can pass through the atrial spetal defect. The stroke volume of the left ventricle is therefore small. Although the arterial oxygen saturation is high, the systemic flow is inadequate. Right ventricular failure develops, the pulmonary flow then decreases, and arterial oxygen saturation falls The pressure rises in both the pulmonary and the systemic veins, and peripheral edema as well as pulmonary edema appears. This state rapidly leads to death

Four of our 11 patients had this severe course, with a fatal outcome during the first two years of life The heart disease was detected soon after birth, because of a murmur Poor weight gain, dyspnea, increased fatigability, slight cyanosis on exertion, and repeated respiratory tract infections were the main clinical features Edema developed a few weeks before death. One patient was operated on at the age of 6 months, when heart failure had just started to develop. She died two days later. The autopsy findings in two of the cases may be inferred from Figures 451b and 453b

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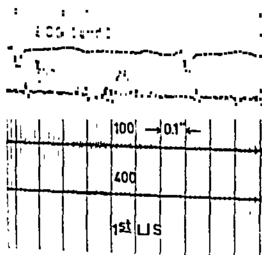


Fig 447.—Phonocardiogram in complete anomalous drainage of pulmonary veins Boy, aged 8 (M.M 460108) A typical continuous murmur is recorded over the first left interspace (L I S) Boxed figures denote degree of amplification, other figures denote standard frequencies of the filters

The *cardiac findings* were characterized by a precordial bulge, parasternal lift, and a systolic murmur over the pulmonary area. The second sound over the pulmonary area was split, and the pulmonary component was accentuated In two cases a venous hum was also audible, it is a frequent occurrence in this malformation (382, 613) Figure 447 shows that the murmur is of the same nature as that in patent ductus arteriosus.

In one patient, an 18-year-old girl, the

physical signs were the same as in pulmonary hypertension. The second sound was greatly accentuated and not split. Over the pulmonary area, only a faint systolic and a protodiastolic murmur were audible. She had been cyanotic since 5 years of age, initially only on exertion, but later at rest as well. Neither cardiac catheterization nor angiocardiography was performed. She died suddenly 18 months after examination. Autopsy showed marked changes in the small pulmonary arteries, with obliteration of the lumen in large areas.

On the *electrocardiogram* the salient features were marked right ventricular hypertrophy, an incomplete right bundle-branch block, and tall P waves over the right precordium (Fig 448)

ROENTGENOLOGIC EXAMINATION

The features common to the different types are due to the left to right shunt. The pulmonary artery is dilated and the vascularity of the lungs is marked, as an expression of the increased blood volume in the pulmonary circulation; the right ventricle and right atrium are enlarged, whereas the aorta is often narrow. The small size of the left atrium is not apparent on the plain roentgenogram. The enlarged right atrium presumably occupies part of the site of the left atrium, so that the decreased volume of the latter chamber is compensated for by this means on the roentgenogram. The heart volume is increased.

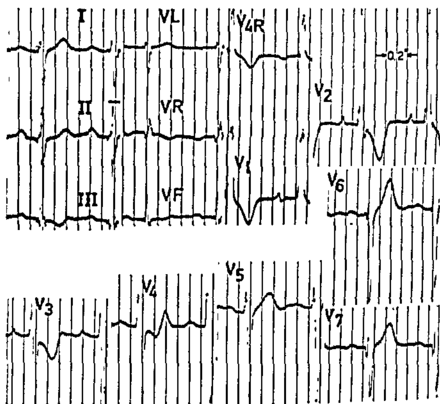


Fig. 448.—Electrocardiogram in complete anomalous venous return. Boy, aged 7 (TA 470220)

1. PULMONARY VEINS OPENING INTO A PERSISTENT LEFT SUPERIOR VENA CAVA— The specific roentgenologic appearance is characterized by great dilatation of the superior part of the mediastinum, caused by the aneurysmally dilated venae cavae (382, 650, 697). The veins are widened, particularly anteriorly and *laterally*, and the dilatation is usually fairly symmetrical in the frontal view (Figs. 449 and 450). The expansion of the veins may occasionally be more pronounced on the left side. Keith

A

B

C

[illegible]

atrium

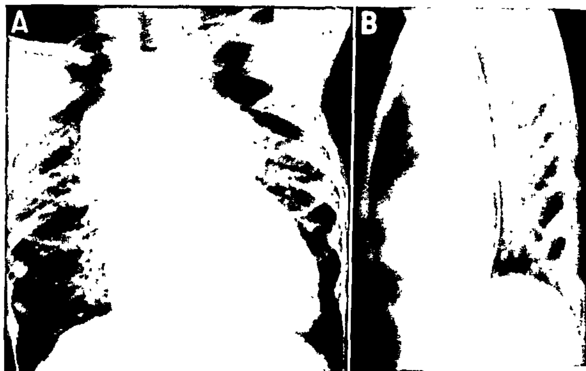


Fig. 450.—Complete anomalous venous return via left superior vena cava and valvular pulmonary stenosis. Boy, aged 7 (T.A. 470220), see Figure 457. Great dilatation of left and right superior venae cavae, enlargement of right atrium and ventricle without signs of hypertrophy, apex rounded but not upturned, greatly increased vascularity of lungs; no enlargement of left atrium; moderate increase in heart volume.

et al. (382) have pointed out that these typical roentgenologic features may be lacking during the first months of life.

In six of our seven cases (Figs 449–452) the roentgenologic appearance was characteristic in every respect. The dilatation of the venae cavae was so great that the aortic arch and pulmonary artery could not be visualized without overlapping. The pulsations in the venae cavae were small Otherwise, all those features characteristic of a left to right shunt at the atrial level were [illegible]

age of 1 year (Figs. 451a and 452). In the remaining case the diagnosis could be established only at autopsy, at 18 months of age.

The abnormal course of the pulmonary veins and the presence of a common venous trunk draining into the vena cava could not be demonstrated on plain roentgenograms, but only with the help of angiocardiography

Blumenthal *et al* (76) found, at repeated examinations of patients with various forms of complete abnormal venous return, that the heart had the normal size at birth and underwent successive, marked enlargement during the following months, with a coincident increase in vascularity of the lungs, which was reminiscent of the conditions in transposition of the great vessels, a typical feature in some of our cases

2. PULMONARY VEINS OPENING INTO THE CORONARY SINUS —Taussig (650) has described the roentgenologic findings in this anomaly Snellen and Albers (613) mentioned a case, verified at autopsy, with an identical roentgenologic appearance In two of our cases, the diagnosis was established post mortem (Figs 453b and 454). All the pulmonary veins opened into the lower part of *the left* superior cardinal vein, corresponding *to the oblique* vein of Marshall and the coronary sinus (Fig. 455). These veins, *which*, even normally, run partly in the posterior wall of the left atrium (see Fig 3a, p 4), partly behind the auricular appendage and partly along the atrioventricular border, were enormously dilated. It may be inferred from Figure 453a that this dilatation, present in one of the cases, simulated enlargement of the left atrium and its appendage, whereas, in reality, the atrium was small. The right *atrium* and ventricle were greatly dilated, as were the pulmonary artery and its central and peripheral branches The shape of the aorta was normal. In Taussig's case, as in our other two cases, no corresponding *dilatation* at the site of the left atrium was seen on roentgenologic examination.

3 PULMONARY VEINS OPENING DIRECTLY INTO THE RIGHT ATRIUM —The roentgenologic appearance was in agreement with that in a large atrial septal defect (Fig. 456).

ELECTROKYMOGRAPHY

See page 515

CARDIAC CATHETERIZATION

In this condition, the anatomy can often be established by means of passage of the catheter into the abnormal pulmonary veins The other most characteristic finding is the same oxygen content in samples from all the chambers of the heart, the pulmonary artery, and the aorta.

Cardiac catheterization was performed in eight of our cases Anatomic pulmonary stenosis was present in two cases, but a pressure gradient was found in other cases as well (Table 11). This is in agreement with the usual findings in the presence of an increased flow through the pulmonary orifice (see p. 120). The arterial oxygen saturation was lower in the cases with pulmonary stenosis or any marked rise in pulmonary artery pressure. In two cases (R.J. 520127 and L.J. 560707) the pressure in the right atrium was raised (mean pressure 9 mm Hg). Both patients died within a month of the appearance of heart failure.

Swan *et al.* (643) found that the oxygen saturation of the pulmonary artery blood often exceeds that of the systemic blood by a few per cent, indicating a preferential



Fig. 451a.—Complete anomalous venous return via left and right superior venae cavae Boy, aged 1 yr (R J 520127) Typical appearance, with dilatation of left superior vena cava (arrows on right), right superior vena cava (arrow on left), right atrium and ventricle, and greatly increased vascularity of lungs No enlargement of left atrium was seen in lateral projection.

Fig. 451b —Same case as in Figure 451a Greatly enlarged right ventricle occupies entire anterior portion of the heart (A ... and both of its bands and the trabeculae (E ... dor sally (D) Right atrium is greatl ... ung of actual wall muscles (C) A ... ava (cephalic portion of left anteric ... the usual way in front of the great ... nal Probe in B is in right innominate vein Left atrium is small (F), arrow points to small crista septal defect. Pulmonary artery is very wide and the aorta narrow (A) AO, aorta, CSV, crista ... , *LIV* and *RIV*, left and right innominate veins, ... d right ventricles, *MV*, mitral valve; *PA*, pul- ... and, *SVC*, superior vena cava, *TV*, tricuspid valve ⟶



Fig. 451b (legend on facing page)

TABLE 11.—COMPLETE ANOMALOUS DRAINAGE OF PULMONARY VEINS: SYMPTOMS AND MAIN FINDINGS ON CARDIAC CATHETERIZATION IN 8 CASES IN RELATION TO SIZE OF THE INTERATRIAL COMMUNICATION*

Case	Age at Death, Year	Interatrial Communication	$\dot{Q}_p/\dot{Q}_s$†	Pressure, mm Hg				Art O_2 Sat, %	Comment
				RV		PA			
				Syst	End-diast	Syst	Diast		
R J 520127 (boy)	11/12	Small foramen ovale	(1.1)‡	70	9	50	20	PA = 79	Heart failure; cath. 1 mo before death; autopsy
L J. 560707 (girl)	6/12	Small foramen ovale	(2.3)‡	79	9	—	—	RV = 88	Operation in early heart failure; patient died 2 days later; autopsy
T A 470220 (boy)	9	Foramen ovale defect 1.5 cm in diameter	1.0	65	7	33	7	84	Patient died at operation; autopsy; patient severely handicapped, could walk 100 m; never heart failure
M M 460108 (boy)	Alive at 10	Foramen ovale defect, size unknown	1.2	65	7	51	13	81	Patient moderately handicapped, can walk 2 km; never heart failure; operation
M J 461024 (girl)	8	Atrial septal defect	1.7	83	10	32	6	75	Valvular pulmonary stenosis; cyanosis since birth; patient severely handicapped, can walk a few 100 m; never heart failure
G B 441109 (boy)	12	Foramen ovale defect, size unknown	5.1	—	—	—	—	94	Patient moderately handicapped; never heart failure
S J 410426 (boy)	14	Large foramen ovale defect	4.1	43	6	25	6	88	Never heart failure; physical working capacity 50% of normal, after operation normal working capacity
G J 280713 (man)	28	Large foramen ovale defect	3.9	42	1	29	6	93	Never heart failure, physical working capacity 55% of normal; operation

*For abbreviations see Table 1, p 119

†$\dot{Q}_p$ = pulmonary flow, $\dot{Q}_s$ = systemic flow

‡Arterial puncture was not performed. Calculations were made on the assumption of an arterial oxygen saturation of the same level as that in the right side of the heart

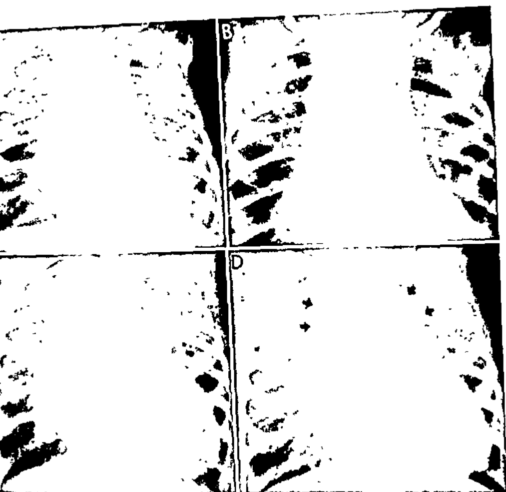


Fig 452 —Complete anomalous venous return Girl, aged 5 months (L J 560707) A–B show the difficulty, during the first six months of life, of visualizing the dilatation of the left superior vena cava, which, together with dilatation of the right superior vena cava, usually gives the picture the characteristic appearance At 1 year of age this dilatation of the vessels is distinct (C–D) Arrows in D point to dilatation of left superior vena cava

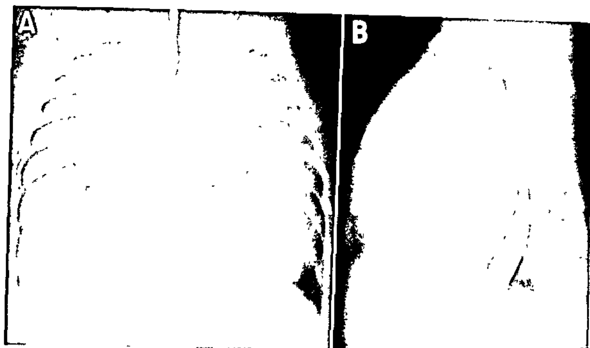


Fig. 453a.—Complete anomalous venous return via coronary sinus. Boy, aged 1 yr (B.L. 530329). Considerable increase in heart volume, enlargement of right atrium and ventricle, greatly increased vascularity of lungs; marked backward bulging of heart at site of left atrium, due to widened coronary sinus.

Fig. 453b.—Same case as in Figure 453a The greatly dilated coronary sinus (*A*), which runs beside the dorsal wall of the small left atrium (*C*), opens into the large right atrium The left atrium and coronary sinus are cut open, and their relative size can be seen *D*, detail of the left atrium Hypertrophy of trabeculae of right atrium Foramen ovale is patent and there are defects in the atrial septum [illegible] ...hy mainly of the trabeculae, but also [illegible] F) runs through [illegible]

the [illegible]
ge.
sinus, *CSV*, crista supraventricularis, [illegible]
nae cavae, *LA*, left atrium, *LAA* and *RAA*,
dge, *PA*, pulmonary artery; *PM*, papillary



Fig 453b (legend on facing page)



Fig. 454.—Complete anomalous venous return through the coronary sinus. Girl, aged 18 (G E 370422). All the pulmonary veins open into the left superior vena cava, which drains via a greatly dilated coronary sinus (*CS*) into the right atrium (*RA*). Left atrium (*LA*) small and [illegible] through a fairly wide septal defect (*ASD*) with the large right atrium High pres- [illegible] of muscles of right ventricle. Pulmonary artery [illegible] branching takes place just distally to the val- [illegible] ventricle



Fig 455 —Complete anomalous venous return, drainage into coronary sinus, wide septal defect, valvular pulmonary stenosis and stenosis at origin of right branch Girl, aged 8 (M.J. 461024), see Figure 460 The appearance is the same as in large atrial septal defect Considerable dilatation of right atrium and ventricle, no dilatation of left atrium, greatly increased vascularity of left lung, normal in right lung

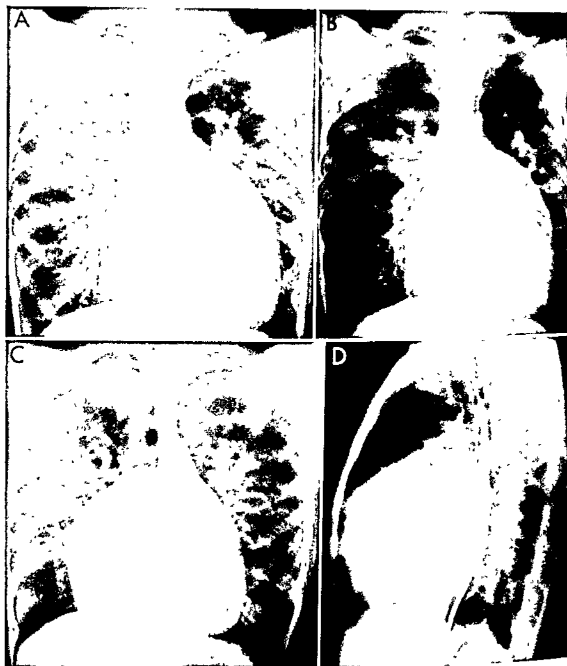


Fig. 456a.—Complete anomalous venous return with great enlargement of right atrium and ventricle Boy, aged 11 (G.B 441109) Greatly increased blood volume in pulmonary circulation, with no enlargement of left atrium

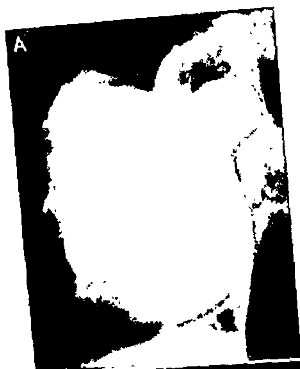


Fig 456b —Same case as in Figure 456a Angiocardiography Right atrium (*RA*) is enormously enlarged, whereas the left atrium (*LA*) is very small consisting chiefly of the appendage

shunting of blood from the inferior vena cava across the atrial septal defect. This is greatly supported by the findings in one of our patients (M J. 461024). In this case all the pulmonary veins opened into the dilated coronary sinus. This emptied into the right atrium and accounted for the left to right shunt. The right to left shunt was through the atrial septal defect. Angiocardiography clearly demonstrated that this shunt was mainly due to the stream from the inferior vena cava, while the blood from the coronary sinus to a high extent entered the tricuspid orifice.

HEMODYNAMICS DURING EXERCISE

In complete anomalous venous return combined with a large interatrial communication, the *hemodynamics at rest* do not differ greatly from those in an isolated large atrial septal defect. In addition to the large left to right shunt there is, however, a small right to left shunt, which is not usually the case in uncomplicated atrial septal defect. The *hemodynamics during exercise* were studied in one adult patient, not included in our material (Table 12). It was found that during exercise the rise in pressure in the pulmonary circuit was greater than normal. The stroke volume of the right ventricle decreased, and the arterial oxygen saturation fell. The systemic flow could not be determined during exercise, but the findings indicate that a relatively greater proportion of the venous reflow passes into the systemic circuit than at rest The systemic arterial pressure can therefore rise in the normal way, but at the cost of decreased arterial oxygen saturation. In our case, the effective stroke volume was only 60 cc at rest In view of the patient's total blood volume (345) it should have been 90 cc.

ANGIOCARDIOGRAPHY

1. PULMONARY VEINS OPENING INTO A PERSISTENT LEFT SUPERIOR VENA CAVA — In this anomaly, the findings on roentgenologic examination and catheterization are so typical that angiocardiography cannot

TABLE 12.—COMPLETE ANOMALOUS VENOUS RETURN*
(Man, aged 28 [G J. 280713], hemodynamic findings at rest and during exercise; body surface area, 1 62 m²; height, 178 cm; weight, 48 8 kg)

Work Load	O_2 Uptake, ml/min	Pulse Rate, beats/min	Pulmonary Circulation			Systemic Circulation			Shunt, L/min		O_2 Cap, vol %	Art O_2 Sat, %	Pressure, mm Hg						
			A-V O_2 diff, ml/L	Cardiac output, L/min	Stroke vol RV, ml	A-V O_2 diff., ml/L	Cardiac output, L/min	Stroke vol LV, ml	L → R	R → L			RA Mean	RV Syst	RV End-diast	PA Syst	PA Diast	Brach art. Syst.	Brach art. Diast
Rest	230	78	10 7	21 5	276	41 8	5 5	70	16 8	0 8	20 5	93	0	42	1	29	6	109	64
200 kg/min	644	106	26.3	24 5	230						20 0	88		71	0	63	18	127	68

*For abbreviations see Table 1, p 119.

be expected to give any information of decisive diagnostic importance. If, however, operation is contemplated, this examination should be made in order to ascertain the anatomic details. From the surgical viewpoint, it is desirable to analyze the position of the pulmonary veins in relation to the left superior vena cava and to the left atrium, respectively, and to determine the size of the left atrium and the width of the mitral orifice.

Angiocardiographic examination was performed in six of our seven cases. In the first three of them, the contrast medium was injected primarily through a catheter with the tip in the caudal part of the aneurysmally dilated left superior vena cava, in the hope of visualizing the pulmonary veins by means of reflux (Figs. 457 and 459). This was not successful, although the injection was made rapidly. The rise in pressure during the injection was insufficient to reverse the shunt. The pulmonary veins could, however, be visualized by means of injection of the contrast medium into the pulmonary artery.

Figures 457–459 show that, in both cases, all the pulmonary veins joined in a single trunk which emptied into the left superior vena cava; it did not communicate with the left atrium. The communication between the left and the right superior vena cava consisted of the greatly dilated left innominate vein.

The left atrium, which was small, was filled from the right atrium, but no detailed conclusions could be drawn regarding the size and position of the septal defect. In three cases (Fig. 457), the left ventricle and the aorta were also visualized. The aorta was narrow, indicating a decreased output to the systemic circulation. The left atrium was distinctly visualized in only two cases. It should be possible to demonstrate the left atrium selectively, by injection of contrast medium through a catheter introduced into the atrium via the atrial septal defect. We have not, however, had reason to perform such a complementary examination.

2. Pulmonary veins opening into the coronary sinus.—In our case, angiocardiography was done on three occasions, with injection into: (1) the right ventricle; (2) the right atrium, the jet of contrast medium being directed toward the atrial septum, and (3) the coronary sinus. All the pulmonary veins opened into the right atrium. The communication between the atria was through an atrial septal defect (Fig. 460).

We also performed angiocardiography in a case of severe tetralogy of Fallot combined with complete anomalous venous return to the coronary sinus disclosed at autopsy. Owing to the markedly decreased blood flow through the lungs, the abnormal venous return could not be visualized angiocardiographically.

3. Pulmonary veins opening directly into the right atrium.—In our case, a diagnosis by exclusion was made on the basis of the findings at catheterization and angiocardiography. The exceedingly small left atrium presupposed the presence of complete anomalous venous return (Fig. 456). At catheterization, the abnormal inflow could be localized to the right atrium.

B. PARTIAL ANOMALOUS DRAINAGE

The anatomic conditions vary greatly. ✓The most usual form is opening of veins from the right upper lobe into the superior vena cava, which was described by Meckel in 1820. The pulmonary veins may also open directly into the right atrium, the inferior vena cava, the azygos vein, the left innominate vein, a persisting left superior vena cava, the left subclavian vein, the coronary sinus, the ductus venosus, the portal vein, or one of the gastric veins (99, 203, 215, 264, 268, 274, 334, 399, 498, 613). These venous anomalies are often associated with severe intracardiac malformations. Here we shall, however, discuss only cases in which the venous anomaly is isolated or the predominant malformation. Atrial septal defects of sinus venosus type usually are associated with anomalous

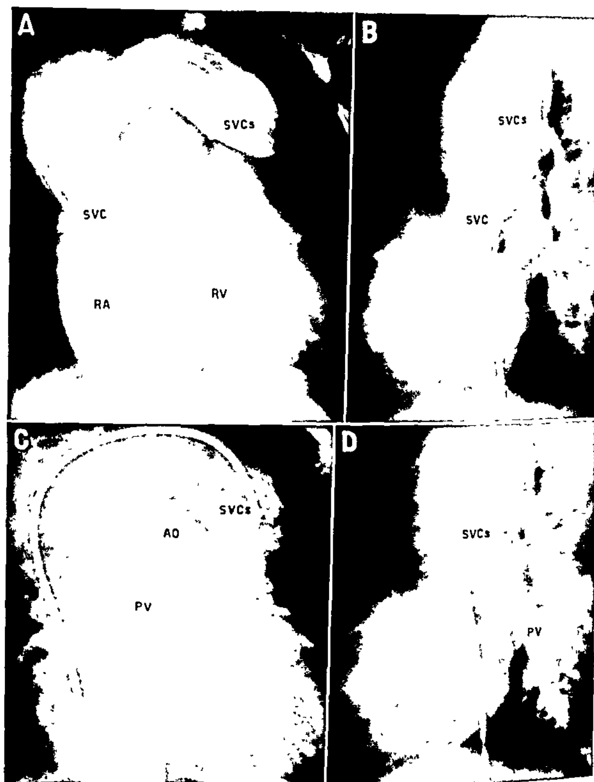


Fig. 457a.—Complete anomalous venous return via left superior vena cava and valvular pul-
... into the pul-
...a cava
...and lies
...n, *RA*,



Fig 457b.—Same case as in Figure 457a Diagram of course of the pulmonary veins and [their] position in relation to the small left atrium *LA*, left atrium

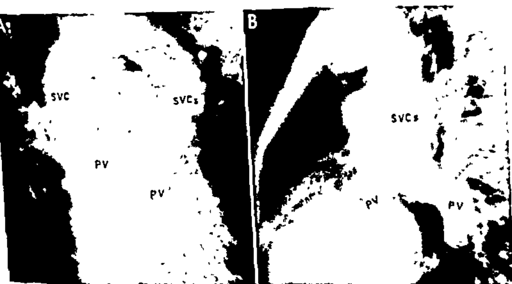


Fig 458.—Complete anomalous venous return Man, aged 28 (G J 280713) All the pulmonary veins (*PV*) open into the left superior vena cava (*SVCs*), which, like the right superior vena cava (*SVC*), is greatly dilated *A*, frontal view, *B*, lateral view

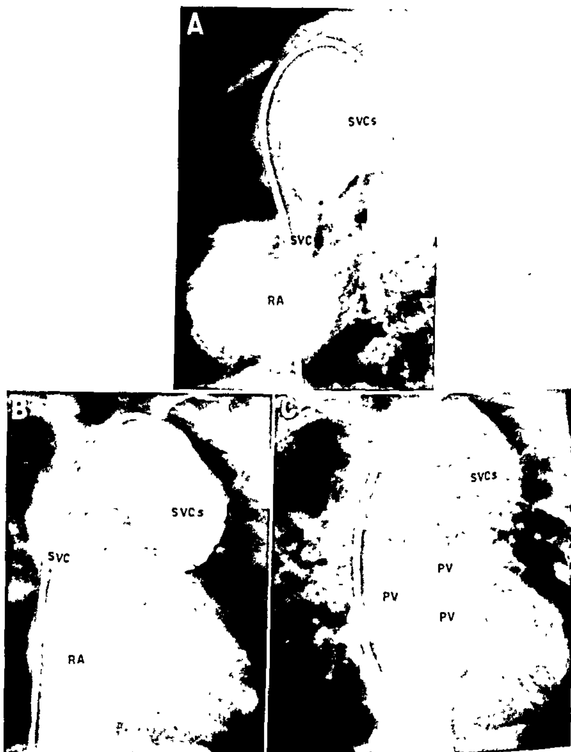


Fig. 459.—Complete anomalous venous return via left superior vena cava. Boy, aged 8 (M M 460108) Contrast medium is injected into the aneurysmally dilated left superior vena cava *A* and *B*, no reflux to the pulmonary veins, but they are filled owing to recirculation They all communicate with the left superior vena cava (*C*) Passage of contrast medium from the right to left atrium is almost inappreciable *PV*, pulmonary vein, *RA*, right atrium, *SVC*, right, and *SVCs*, left, superior vena cava

drainage of the veins from the upper or middle lobe of the lung. These cases have been classified as atrial septal defect

Our series contains five cases A foramen ovale defect was present in two cases, and in one of them there was mild valvular pulmonary stenosis as well. The veins from the right lung or one of its lobes opened into the superior vena cava 1 to 2 cm from its entry into the right atrium In one case a more complicated venous anomaly was present, it is described in greater detail in connection with the angiocardiographic findings (see p 524)

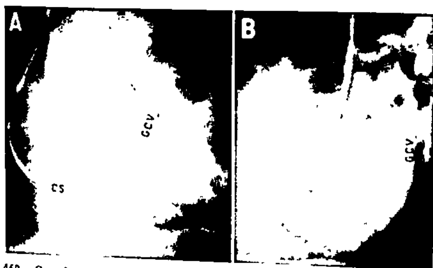


Fig 460 —Complete anomalous venous return, drainage into right atrium through coronary sinus Girl, aged 8 (M J. 461024) Catheter is inserted into the coronar[illegible]
made under low or pre[illegible]
co[illegible]
pe[illegible]

CLINICAL FEATURES

If only half of the pulmonary flow is drained into the right side of the heart, symptoms are seldom apparent in childhood and the patient may live to an advanced age (613) The malformation nevertheless implies an increased pulmonary flow and a resulting increased load on the right side of the heart, although cardiac failure does not develop until adulthood

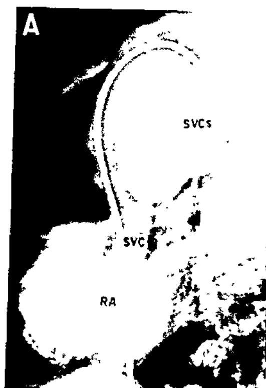
The *cardiac findings* were the same as in moderately severe atrial septal defect, i e., a parasternal lift as an expression of right ventricular enlargement, a systolic murmur, and a split second sound over the pulmonary area (Fig 461).

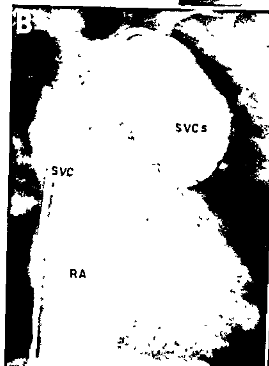
The *electrocardiogram* also exhibited the same changes as in atrial septal defect, namely, an incomplete right bundle-branch block, sometimes combined with right ventricular hypertrophy. Both of these features were present in three cases, whereas in one there was right ventricular hypertrophy only, and in the fifth one there was an incomplete right bundle-branch block alone

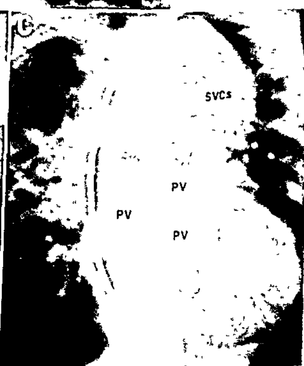
ROENTGENOLOGIC EXAMINATION

As in the preceding group of cases, the common roentgenologic features are dependent on the left to right shunt They are usually considerably less conspicuous than in complete anomalous venous return

1 PULMONARY VEINS OPENING INTO SUPERIOR OR INFERIOR VENA CAVA OR DIRECTLY INTO RIGHT ATRIUM —When the veins of the right lung are drained into the inferior vena cava, this is usually visualized dis-







F - 459 Complete anomalo... 't superior vena cava Boy, aged 8 (M M ... nally dilated left superior vena cava A ... 'd owing to recirculation They all com- ... contrast medium from the right to left ... RA, right atrium, SVC, right, and SVCs, left, superior vena cava

tinctly on ordinary roentgenologic examination The larger pulmonary veins have a normal course paracardially, and usually . . . .

by Dotter, Hardisty, and . . .

left, and anomalies of the right lung are common, in the form of malformed and abnormally distributed bronchi, as well as an abnormal arterial inflow of vessels from the descending thoracic aorta (Halasz *et al*, 313).





Fig 462b.—Same case as in Figure 462a All the pulmonary veins (*PV*) on the right side drain into the superior vena cava and open into its lower part The pulmonary veins on the left side open into the left atrium (*LA*), which is small *AO*, aorta, *PA*, pulmonary artery

Grishman *et al* (302), Arvidsson (21), Findlay and Maier (255), Welti and Nedey (694), Runstrom and Sigroth (572), and Bruwer (114), among others Angiocardiography was performed in most of the cases and the pulmonary veins were visualized in detail In such cases the right hemithorax usually is considerably smaller than the

We investigated a case in which all the veins of the right lung joined a trunk which emptied into the superior vena cava at its entry into the right atrium (Fig 462). The venous trunk in question did not, however, appear as distinctly as in cases with the pulmonary veins opening into the inferior vena cava A remarkable observation in our

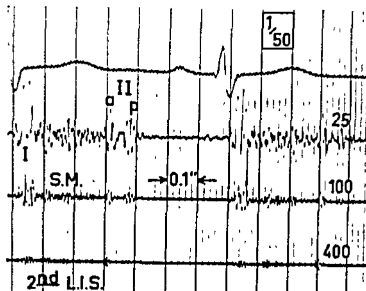


Fig. 461.—Phonocardiogram in case of drainage of veins of the right lung into superior vena cava Girl, aged 18 (I D 360412). Findings are the same as in atrial septal defect: a fairly faint systolic murmur (*SM*) and widely split but not accentuated second sound. Boxed figures denote degree of amplification; other figures denote standard frequencies of the filters. *II a* and *p*, aortic and pulmonary components of the 2nd sound; *L.I.S.*, left interspace.



Fig. 462a.—*Partial anomalous venous return* Girl, aged 15 (A O. 411125) The pulmonary veins on the right side have an abnormal course, so that the vascularity is sparse paracardially (*B*) All the pulmonary veins run toward the superior vena cava.

tinctly on ordinary roentgenologic examination The larger pulmonary veins have a ...al course paracardially, and usually ...

by Dotter, Hardisty, and ...

left, and anomalies of the right lung are common, in the form of malformed and abnormally distributed bronchi, as well as an abnormal arterial inflow of vessels from the descending thoracic aorta (Halasz et *al*., 313).





Fig 462b —Same case as in Figure 462a All the pulmonary veins (*PV*) on the right side drain into the superior vena cava and open into its lower part The pulmonary veins on the left side open into the left atrium (*LA*), which is small *AO*, aorta, *PA*, pulmonary artery

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Fig. 463.—Partial anomalous venous return and atrial septal defect of sinus venosus type. Woman, aged 24 (G E 290918). Considerable enlargement of right atrium and ventricle, without signs [illegible] a lengthy contiguity to the anterior thoracic [illegible] r part of the superior vena cava (*C*). The vess[illegible] which the catheter lies in *C* It runs cephalad, in an abnormal direction, and is not given off by the pulmonary artery Considerably increased vascularity of lungs, pulmonary artery wide, no enlargement of left atrium

case was the sparse vascularity paracardially on the right side. The lung was not underdeveloped, and the right and left hemithorax were symmetrical.

If, on the contrary, isolated pulmonary veins open into the superior vena cava or directly into the right atrium, the roentgenologic appearance is more difficult to analyze The features usually give no reason for one to suspect the presence of anomalous venous return, but are in every respect in conformity with the appearance in uncomplicated atrial septal defect.

In *our series*, pulmonary veins opening into the superior vena cava or into the right atrium were identified by other means in five cases In three of them, an associated atrial septal defect could be identified with certainty. In the other two, the presence of such a defect could be ruled out with a high degree of probability The difficulty of demonstrating on the roentgenograms that the pulmonary veins have an aberrant opening seems to be dependent mainly on the fact that the course of the veins differs to such a slight extent from the normal course. In one case it could, however, be shown that the veins joined into *one* trunk instead of into two, as normally, and that the venous trunk lay strikingly far cephalad. This indicated an abnormal course of the *inferior* pulmonary vein on the right side (Fig 464) A pulmonary vein opening into the right atrium was disclosed in two cases by direct passage of the catheter from the atrium into the vein (Fig 465)

Figure 463 shows a pulmonary *vein* which had a course entirely in agreement with that which could be catheterized from the superior vena cava. Presumably, the vein drained only a small part of the lung. The greatly increased blood volume in the pulmonary circulation was due essentially to a large atrial septal defect

2 Pulmonary veins opening into the azygos vein —We have one such case in our series. The course of the azygos vein was abnormal, and the peculiar anatomy is evident from Figure 466

3 Pulmonary veins opening into the innominate vein —Patterson (529) described a case in which the veins of the upper lobe and the superior part of the lower lobe on the left side drained into the left innominate vein. We have no such case in our series.

ELECTROKYMOGRAPHY

In six of the 10 cases studied, the electrokymograms from the pulmonary artery presented the same characteristic change in shape as in uncomplicated atrial septal defect (Figs. 467 and 468).

The tracings from the right atrium were normal except in one case In this case the electrokymogram was typical of an impediment to atrial emptying (Fig. 469). One variant is illustrated in Figure 468 Curves were also recorded over the dilated superior venae cavae in three cases in which there was complete anomalous venous return (Fig. 470). Electrokymography confirmed that the pulsations were of venous type.

CARDIAC CATHETERIZATION

If the aberrant pulmonary veins open directly into the right atrium, the findings on catheterization are the same as in atrial septal defect. As a rule, however, they open into the superior vena cava This anomaly can be distinguished from atrial septal defect by taking several samples at different *levels* of the superior vena cava The most certain means of demonstrating the anomaly is by passing the catheter into the relevant pulmonary vein from the superior vena cava This could be done in all our cases On the other hand, it is exceedingly difficult to determine whether the catheter *has passed* from the right atrium directly into the pulmonary vein, or whether it has first passed through the foramen ovale and the left atrium It may also be difficult to determine, on the basis of passage of the catheter into the pulmonary veins, how many lobes have anomalous drainage We therefore performed *angiocardiographic* examination to obtain a more detailed anatomic diagnosis (see p 522). If the catheter passes into the left atrium, this does not

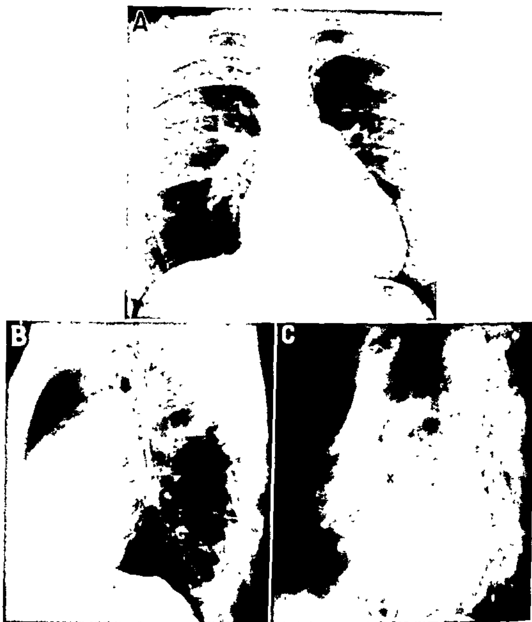


Fig. 464a.—Partial anomalous venous return and atrial septal defect of sinus venosus type Girl, aged 18 (B L. 360118) Moderately increased heart volume, enlargement of right atrium and ventricle The ventricular surface has a long contiguity to the anterior thoracic wall, as in dilatation. Moderately increased vascularity of lungs Pulmonary veins on the right side join in a thick trunk, at an abnormally high level (x in C) No enlargement of left atrium Aorta narrower than normally

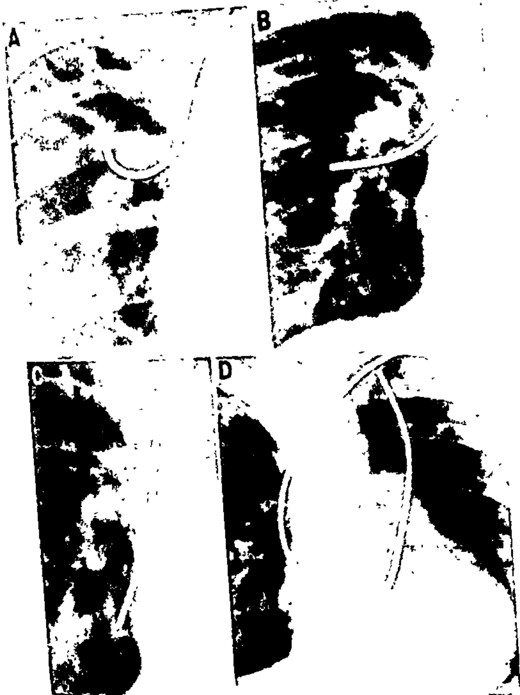


Fig 464b —Same case as in Figure 464a *A–C*, catheter is inserted in different pulmonary veins on the right side The superior veins open into the lower part of the superior vena cava (*A* and B), the inferior veins open into the sinus venarum (*C*) *D*, catheter is advanced through the atrial septal defect into one of the superior pulmonary veins on the left side



Fig. 465.—Partial anomalous venous return and patent foramen ovale (see Fig 474, p 525) Girl, aged 8 (I.L 471116). The catheter from the left arm passes via the right atrium into a pulmonary vein with an anomalous opening The tip of the catheter introduced from the right leg lies in the left atrium.

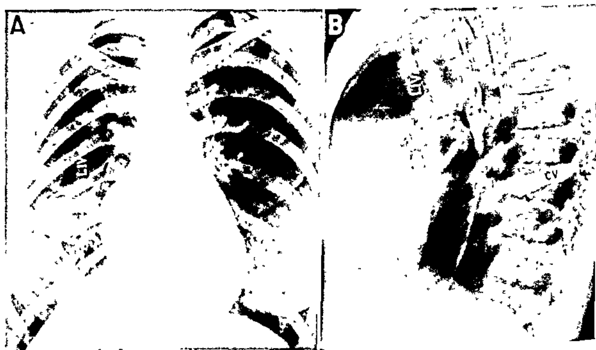


Fig. 466a—Partial anomalous venous return and abnormal systemic veins Woman, aged 26 (M L 270630), see Figure 475 Left innominate vein, which lies far out in the paren- ... into the



Fig 466b.—Same case as in Figure 466a Catheter inserted in left innominate vein (C and *D*), it could be inserted into the right atrium via the azygos vein emptying at a low level (C) as well as into a pulmonary vein (*D*) *A* and *B*, catheter inserted in posterior cardinal vein (azygos vein) Inferior vena cava is lacking Azygos vein joins the left innominate vein in a common trunk Catheter tip lies in right branch of pulmonary artery. *E*, catheter inserted in right innominate vein *CV*, right posterior cardinal vein, *LIV* and *RIV*, left and right innominate veins, *PA*, pulmonary artery

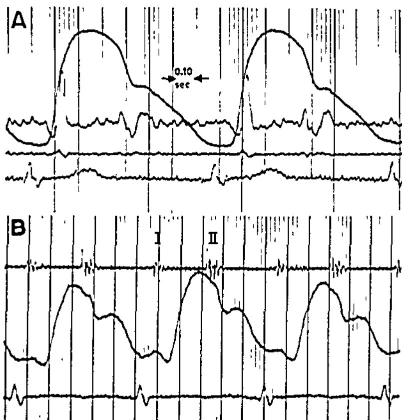


Fig. 467.—Electrokymograms of pulmonary artery in partial anomalous venous return and atrial septal defect. Woman, aged 26 (M L. 270630). Frequency channels, 10 and 20 cps Time marking, 0.10 sec between thick lines. *A*, before operation The part of the curve corresponding to reduced ejection is broad and rounded Protodiastole, 0.06 sec. The dicrotic wave is low. Pressure in RV 30/3 mm Hg, in PA 14/4 *B*, after operation PCG over pulmonary area *I*, 1st sound, *II*, 2nd sound The electrokymogram has normal appearance Pressure: in RV 17/4 mm Hg, in PA 15/4

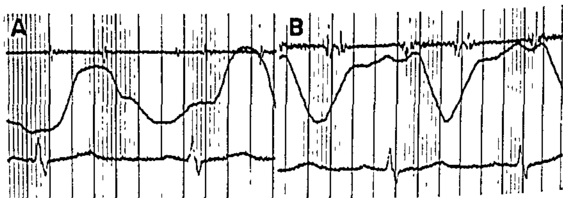
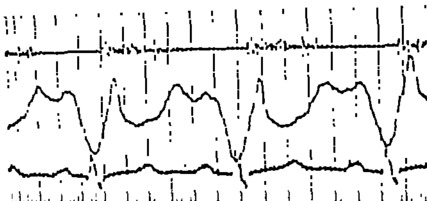


Fig. 468.—Electrokymograms in partial anomalous venous return Boy, aged 6 (A B 470612) *A*, pulmonary artery PCG over 3rd left interspace Tracing has the same appearance as in Figure 467, *A*. *B*, right atrium PCG over pulmonary area Deep systolic deflection owing to influence of right ventricle. Emptying phases in presystole and systole are shallow Configuration of the curve is not pathologic



469 —Electrokymogram of right atrium in complete anomalous venous return Boy, aged
J 410426) PCG over 2nd L I S The deflection during atrial systole has a duration of 0 10
d a greater relative depth than the deflection in the early part of diastole. The curve has
pearance typical of an impediment to atrial emptying

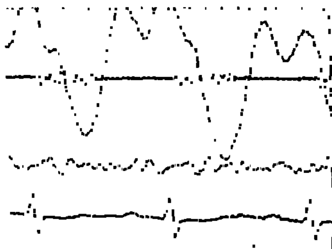


Fig 470 —Electrokymogram of dilated left superior vena cava in complete anomalous venous
rn Boy, aged 11 (T.A 470220) PCG over pulmonary area Electrokymogram shows venous
sations

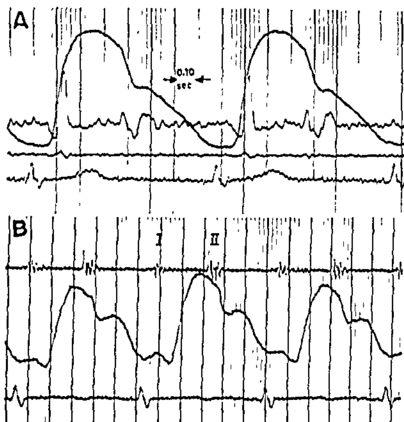


Fig. 467.—Electrokymograms of pulmonary artery in partial anomalous venous return and *atrial septal defect* Woman, aged 26 (M.L 270630). Frequency channels, 10 and 20 cps Time marking, 0 10 sec between thick lines *A*, before operation. The part of the curve corre- [illegible] Protodiastole, 0 06 sec The dicrotic wave is [illegible] after operation PCG over pulmonary area I, [illegible] is normal appearance Pressure. in RV 17/4 mm Hg, in PA 15/4

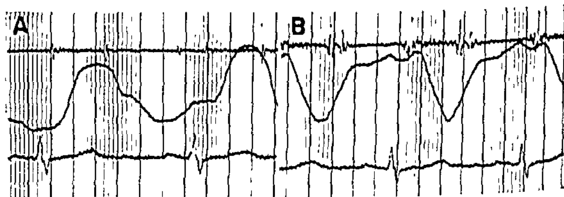


Fig. 468.—Electrokymograms in partial anomalous venous return Boy, aged 6 (A B 470612). *A*, pul[illegible] the same appearance as *in Figure 467, A* [illegible] o in- fluence of right [illegible] on of the curve is not pathologic.

view, since the operative procedure differs according to whether there is anomalous venous return only, an atrial septal defect, or a combination of both malformations.

The contrast medium can be injected either into the main trunk of the pulmonary artery or into the right ventricle. Despite this selective procedure, the density of the contrast medium is definitely decreased in the pulmonary veins, owing to great dilution. In some of our cases opacification oc-



Fig 472 —Partial anomalous venous return to right superior vena cava and superior part of sinus venarum Girl, aged 18 (I D 360412) Left catheter (*1*) lies in the pulmonary vein, opening into the superior vena cava, right catheter (*2*) passes through the right atrium into a pulmonary vein which opens into the superoposterior part of the sinus venarum

curred simultaneously in the two atria, in the left it was seen earliest in the left lateral segment, and in the right in the superior segment, thus indicating that a pulmonary vein emptied into the right atrium In the lower part of this chamber there was, on the contrary, distinct dilution caused by the flow from the inferior vena cava

An aberrant pulmonary vein can be visualized selectively by injection of the contrast medium via a catheter introduced into the vein (Tori, 654) It is important that the tip of the catheter should not obliterate the vessel and that the quantity of contrast medium be sufficient to permit visualization of both the pulmonary vein and its connection with the inflow tract of the atrium. If there is reason to suppose that several pulmonary veins are drained anomalously, this technique requires visualization of each of them separately.

On rapid injection of contrast medium into the vena cava, transient retrograde filling of a pulmonary vein opening into it is sometimes obtained Nordenström has shown that if the intrathoracic pressure is temporarily greatly increased, this leads to considerably better opacification of such pulmonary veins (Fig. 473). In peripheral (venous) angiocardiography, the contrary, no reflux to pulmonary veins can be anticipated The appearance of local dilution effects in the vena cava or right atrium can, however, indicate the presence of abnormal emptying in such cases (254, 596).

necessarily imply the presence of an atrial septal defect, but there may be a patent foramen ovale in combination with anomalous pulmonary veins. This can be elucidated with the help of a balloon catheter (see p. 445) or by angiocardiography with injection into the left atrium. The possibility of differentiating between atrial septal defect and anomalous pulmonary venous connection by means of indicator-dilution techniques has been discussed in a series of papers from the Mayo Clinic (637, 640, 642, 644).

All of our cases had normal pressure in the pulmonary artery. Mild valvular pulmonary stenosis was present in one case. The

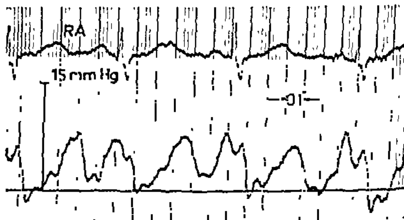


Fig. ... from right atrium Boy, aged 7 (A B ... s not raised. ...

systolic pressure in the right ventricle was 50 mm Hg and in the pulmonary artery 25 mm In all but one case the catheter advanced into the left atrium. In two cases there was only a patent foramen ovale, and in two a small atrial defect was present. No right to left shunt could be demonstrated *The pressure in the left atrium was higher* than in the right, but the difference was not as great as normally, amounting to only 1 to 3 mm Hg The pressure in the right atrium was not, however, significantly raised (Fig. 471). Owing to the increased flow through the right atrium, a rise in pressure would have been anticipated, particularly during ventricular systole. It is true that the *v* wave was as high as the *a* wave, but it was not abnormally high. Since the right atrium is distensible, it permits a large flow without a rise in pressure.

ANGIOCARDIOGRAPHY

1. PULMONARY VEINS OPENING INTO THE INFERIOR VENA CAVA.—In several of the cases reported in the literature, the abnormal vascular anatomy of the right lung was visualized on angiocardiography. We have no case of our own to illustrate this fact

2. PULMONARY VEINS OPENING INTO RIGHT SUPERIOR VENA CAVA AND RIGHT ATRIUM.—One or several pulmonary veins emptying into the right superior vena cava immediately above the orifice of the right atrium can be identified on cardiac catheterization (Figs. 463, 464, and 472). As already stated, if the catheter passes into a pulmonary vein after being introduced into the right atrium, it may be difficult or impossible to determine whether it has also passed through the atrial septum. This applies particularly when the catheter has been introduced through a leg vein Consequently, it is often uncertain whether the pulmonary vein opens into the right or the left atrium.

Angiocardiography is then warranted in order, if possible, to ascertain the anatomy and course of the pulmonary veins. This is of importance from the surgical point of



Fig 474 —Partial anomalous venous return and patent foramen ovale Girl aged 8 (I L. 471116). On injection into the l... right atr... the valve A-... f the

It should be possible to obtain additional information by blocking one of the branches of the pulmonary artery with a balloon and making the injection through the catheter peripherally to the balloon, according to Nordenström's technique (506).

An eventual concomitant atrial septal defect can be studied after introduction of the balloon catheter into the left atrium, or by rapid injection of contrast medium into this chamber (p. 450). If the defect is blocked even on slight distention of the balloon, or if there is little or no passage of contrast medium into the right atrium, a patent foramen ovale presumably is present (Fig. 474). *In this event, aberrant pulmonary veins are entirely responsible for the shunts, provided that the presence of a communication between the left atrium and coronary sinus can be ruled out (p. 486)*

3. PULMONARY VEINS OPENING INTO THE AZYGOS VEIN.—In our case, the object of angiocardiographic examination was to identify the normal and pathologic pulmonary veins and to visualize the abnormal *flow into the affected systemic veins.*

After the catheter had been introduced from the azygos vein, via the right atrium, into the right ventricle, the dilated pulmonary artery and its branches could be visualized selectively (Fig. 475). In the right lung, the branches to the lower and middle lobes were somewhat elongated. The vessels were nevertheless of the same width as those of the left lung, and no hypoplasia of the vascular tree of the kind observed in cases with anomalous venous return to the inferior vena cava (203) was present. The aberrant pulmonary veins emptied into the azygos vein. The other pulmonary veins opened into the left atrium

The hepatic veins were visualized by

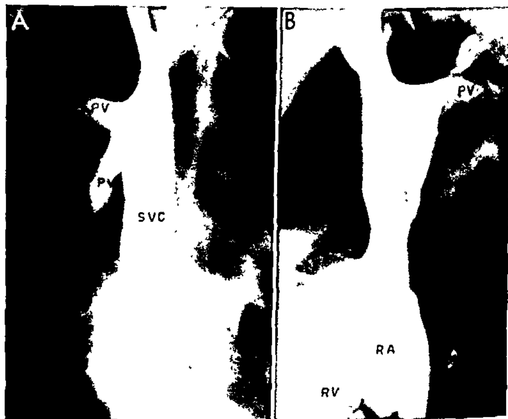


Fig. 473.—Partial anomalous venous return and coarctation of the aorta. Girl, aged 16 (E I. 410918) The veins of the right upper and middle lobe (*PV*) open posteriorly into the superior ... vena ... esy of

... the right atrium Car-

4 PULMONARY VEINS CONNECTING WITH LEFT SUPERIOR VENA CAVA, DRAINING INTO THE RIGHT ATRIUM AND ALSO CONNECTED TO THE RIGHT SUPERIOR VENA CAVA — Roentgenologic findings in such cases have been described by Fitzgerald Peel *et al.*

(135). In our series, this anomaly was observed in, for example, ventricular and atrial septal defect, tetralogy of Fallot, mitral atresia, and the Taussig-Bing complex. It can almost invariably be demonstrated on ordinary roentgenologic examination except in cases in which the mediastinal vessels are masked by the thymus. The course of the vessel in the lateral, left superior part of the mediastinum is typical (Fig 476).



Fig. 476.—Persisting left superior vena cava and mitral atresia Girl, aged 1 yr (S A. 511101) Right superior vena cava is probably lacking The azygos vein (arrow) opens into the left superior vena cava (SVCs)

(256). Similar reports of drainage of a left superior pulmonary vein into the left superior vena cava have been given by Ödman (515), and of total drainage of pulmonary veins into the right superior vena cava and right atrium by Sepulveda *et al* (596), among others We have seen none of these types

ANOMALOUS SYSTEMIC VEINS WITHOUT A RIGHT TO LEFT SHUNT

A persistent left superior vena cava is a fairly common secondary finding in the investigation of congenital heart disease

The anomaly may appear as an isolated malformation. The additional details of the course are best studied by cardiac catheterization or intravenous angiocardiography. The vein usually opens into the coronary sinus (Fig. 476).

Anomalies of the azygos and the hemiazygos vein are illustrated in Figures 8 (p. 10), 477, and 478 In one case (Fig 478) the normal communication between the azygos and the hemiazygos vein had not developed The hemiazygos vein was drained instead into the left innominate vein through the accessory hemiazygos vein. Since the thymus was at an abnormal site

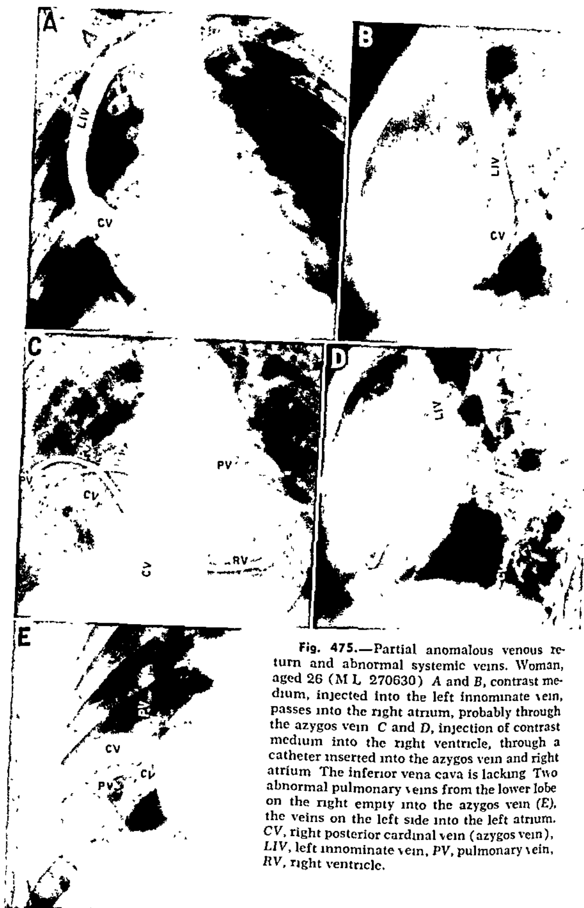


Fig. 475.—Partial anomalous venous return and abnormal systemic veins. Woman, aged 26 (M L 270630) *A* and *B*, contrast medium, injected into the left innominate vein, passes into the right atrium, probably through the azygos vein *C* and *D*, injection of contrast medium into the right ventricle, through a catheter inserted into the azygos vein and right atrium The inferior vena cava is lacking Two abnormal pulmonary veins from the lower lobe on the right empty into the azygos vein (*E*), the veins on the left side into the left atrium. *CV*, right posterior cardinal vein (azygos vein), *LIV*, left innominate vein, *PV*, pulmonary vein, *RV*, right ventricle.

means of reflux from the right atrium. Cardiac catheterization, angiocardiography, and operation showed that the inferior vena cava was lacking

4 PULMONARY VEINS CONNECTING WITH A LEFT SUPERIOR VENA CAVA, DRAINING INTO THE RIGHT ATRIUM AND ALSO CONNECTED TO THE RIGHT SUPERIOR VENA CAVA.— Roentgenologic findings in such cases have been described by Fitzgerald Peel *et al.* (135). In our series, this anomaly was observed in, for example, ventricular and atrial septal defect, tetralogy of Fallot, mitral atresia, and the Taussig Bing complex. It can almost invariably be demonstrated on ordinary roentgenologic examination except in cases in which the mediastinal vessels are masked by the thymus. The course of the vessel in the lateral, left superior part of the mediastinum is typical (Fig 476).



Fig 476 —Pe... Right superior vena cava (SV...

(256) Similar reports of drainage of a left superior pulmonary vein into the left superior vena cava have been given by Ödman (515), and of total drainage of pulmonary veins into the right superior vena cava and right atrium by Sepulveda *et al* (596), among others We have seen none of these types

ANOMALOUS SYSTEMIC VEINS WITHOUT A RIGHT TO LEFT SHUNT

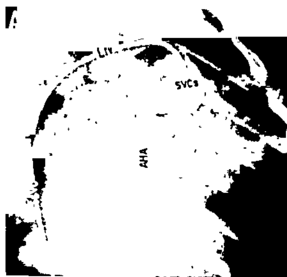
A persistent left superior vena cava is a fairly common secondary finding in the investigation of congenital heart disease

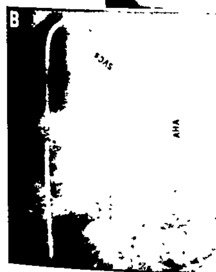
The anomaly may appear as an isolated *malformation*. The additional details of the course are best studied by cardiac catheterization or intravenous angiocardiography. The vein usually opens into the coronary sinus (Fig 476).

Anomalies of the azygos and the hemiazygos vein are illustrated in Figures 8 (p. 10), 477, and 478. In one case (Fig. 478) the normal communication between the azygos and the hem... veloped T... instead i... innominate vein through the accessory hemiazygos vein. Since the thymus was at an abnormal site



Fig. 477.—Abnormally draining systemic veins Boy, aged 7 (A R. 470325); see Figure 478 Increase in breadth of superior part of the mediastinum, somewhat reminiscent of complete anomalous venous return to the left superior vena cava. It does not have the same rounded appearance. Catheter is passed into the right superior vena cava and left innominate vein Tip has advanced into the left superior vena cava, which drains the hemiazygos vein and probably the [illegible] bronchus, at the azygo [illegible] the thylevel [illegible] ly. mus,





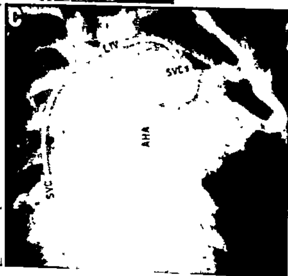


Fig 478 —Anomalous drainage of systemic veins same
dium, injected
part of the
empties in
left superi
inate vein, SVC, ri

to the left in the mediastinum, the appearance bore a faint likeness to that in anomalous venous return to a persisting left superior vena cava. The shape of the heart and the vascularity of the lungs had, however, the ordinary appearance A similar malformation was described by Weber (687) in 1829

The other case (Fig 8), in which more comprehensive anomalies of the systemic veins were present, is described in detail in Chapter 1, on Embryology

In our series, we had no cases in which systemic veins opening into the left atrium gave rise to cyanosis (256, 267, 353, 452, 658)



Fig. 477.—Abnormally draining systemic veins Boy, aged 7 (A.R 470325), see Figure 478 Increase in breadth of superior part of the mediastinum, somewhat reminiscent of complete anomalous venous return to the left superior vena cava It does not have the same rounded appearance. Catheter is passed into the right superior vena cava and left innominate vein Tip has advanced into the left superior vena cava, which drains the hemiazygos vein and probably the azygos vein No vessel visible at the site of the azygos vein above the right main bronchus, at the level of the bifurcation (arrow in *D*) In *A*, the triangular bulge (arrow) is caused by the thymus, which lies farther to the left than normally, probably owing to the vascular anomaly.

are of interest in a discussion of this question

BOY, PREMATURE (G L 490812).—The infant weighed 1,020 Gm at birth During his first three weeks of life, no murmur was audi-

per minute. He resumed breathing in a few minutes, and a continuous murmur typical of patent ductus was then heard instead The same observations were made during several attacks of asphyxia which occurred in the following weeks The auscultatory findings were subsequently entirely normal The latest examination was made when he was 2 years old, and the heart findings were then normal.

GIRL, NEWBORN (L C 520611) —The patient had hemolytic disease of the newborn She exhibited signs of cerebral involvement

and her

was performed at the age of 4 weeks (Table 13)

On this occasion there was a continuous murmur, and gas analysis showed a left to right shunt to the pulmonary artery The right ventricular pressure was probably normal (normal values for this age group are lacking) On the patient's breathing 100 per cent oxygen the shunt decreased, since the pressure remained unchanged, this must have been due to narrowing of the ductus arteriosus When the patient was discharged from the hospital at the age of 2 months, the murmur was still audible When she

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BOY, NEWBORN (G.J 521011) —The infant was transferred from the obstetric clinic on his first day of life because of hemolytic disease of the newborn His respiration was extremely irregular and he was slightly cyanotic. During the first three weeks, a systolic murmur of varying intensity was heard over the pulmonary area The auscultatory findings were subsequently normal He was given repeated blood transfusions, but slight anemia persisted Dur-

tion of the respiration and an improvement in the general condition At 15 days of age, cardiac catheterization was performed in association with an exchange transfusion. The catheter was introduced with the utmost caution into the pulmonary artery, it passed with ease through the ductus arteriosus into the descending aorta The relevant pressures and results of the gas analyses are shown in Table 13 The fact that the ductus was patent was demonstrated by the passage of the catheter. The difference between the oxygen content of the right ventricle and that of the pulmonary artery was not sufficient to afford definite evidence of a shunt The most probable explanation of the murmur heard in the first three weeks of life is the patency of the ductus, the findings on cardiac catheterization give some support to this assumption

TABLE 13 —MAIN FINDINGS ON CARDIAC CATHETERIZATION IN 2 CASES OF PATENT DUCTUS ARTERIOSUS IN THE NEONATAL PERIOD*

Case	Pressure mm Hg			O_2 Content vol %		Comments
	RV	PA	Aorta	RV	PA	
L C. 520611 (4 weeks)	30/0	—	—	9 2	11 1	Air breathing
	30/3	30/18	—	10 0	10 4	100% O_2 breathing
G J 521011 (15 days)	33/0	37/22	70 60	9 8	10 4	Air breathing

*The ductus subsequently closed spontaneously In case L C 520611 the shunt decreased on breathing 100 per cent oxygen

Both the clinical observations and the results of cardiac catheterization indicate that asphyxia retards closure of a normal ductus arteriosus On the other hand, it is not known whether this condition is in any

follow-up study of the mental and physical development in 85 children with asphyxia neonatorum made by one of us in collaboration with Hellstrom, no case with a patent ductus was found (328)

ANATOMY —The ductus arteriosus varies greatly in width, length and shape. It is

16 Patent Ductus Arteriosus

WHEN A PATENT ductus arteriosus occurs in combination with an intracardiac malformation or an anomaly of the aortic arch system, it may have a beneficial effect on the circulation. Conversely, as an isolated lesion it always implies an increased burden on the heart. In all probability it is a true malformation and not merely a persistence of a communication normally present during fetal life. When an extremely wide ductus is found in newborn infants, there is an unquestionable malformation of the vessel which has prevented its closure at birth. Anatomic changes have also been found in the presence of a small patent ductus. Thus, the wall—particularly the intima—is far thinner than normally and the media contains much collagen (216)

CLOSURE OF THE NORMAL DUCTUS ARTERIOSUS.—The time at which the ductus normally closes has been much discussed. It is essential in this connection to distinguish between the anatomic and the functional closure. Complete obliteration takes place at a greatly varying time, ranging from two weeks to several months after birth (154). Functional closure is presumably brought about by contraction of the muscles of the duct. The wall of the ductus arteriosus is more muscular than that of other arteries (645). Barcroft's (39) physiologic studies of the fetal and neonatal circulation in the lamb showed that the ductus ceases to function a few minutes after birth. Dawes and his co-workers (183) reported similar experiments, but found that the ductus was still patent 1½ hours after the beginning of ventilation. Experimental studies have shown that an increase in arterial oxygen tension promotes closure of the ductus (81, 384, 550).

Eldridge and Hultgren (230) found lower oxygen saturation of the arterial blood in the foot than in the hand in many healthy infants during the first days of life This might have been due to a veno-arterial shunt through the ductus arteriosus. However, the babies were crying during sampling of the blood, and, in our experience, the pressure in the pulmonary artery can then rise to up to three times the resting level. The angiocardiographic technique has also been used to study closure of the ductus in infants (379, 437). The contrast medium may, however, affect vascular tonus (655) and thereby bring about a change in pressure in both the systemic and the pulmonary circuits. For this reason, it is difficult to draw any definite conclusions regarding the size and direction of the shunt

It is a common observation that a systolic murmur may appear during the first days of life and then disappear entirely. It has been assumed that this is caused by functioning of the ductus arteriosus, which then closes after a few days (650). No proof has been brought forward in support of this hypothesis. The following cases, which we had the opportunity of observing.

re of interest in a discussion of this question

Boy, premature (G L. 490812) —The infant weighed 1,020 Gm at birth During his

[illegible]

per minute He resumed breathing in a few minutes, and a continuous murmur typical of patent ductus was then heard instead The same observations were made during several attacks of asphyxia which occurred in the following weeks The auscultatory findings were subsequently [illegible]

[illegible]

she exhibited signs of cerebral involvement and her respiration was slow and deep The hemolytic anemia was corrected by means of repeated blood [illegible]

[illegible]

On this occasion there was a continuous murmur, and gas analysis showed a left to right shunt to the pulmonary artery The right ventricular pressure was probably normal (normal values for this age group are lacking) On the patient's breathing 100 per cent oxygen, the shunt decreased, since the pressure remained unchanged, this must have been due to narrowing of the ductus arteriosus When the patient was discharged from the hospital at the age of 2 months, the murmur was still audible When she was readmitted two months later because of severe brain damage, no pathologic murmur could be heard She died at the age of 8 months No cardiac malformation was found at autopsy and the ductus arteriosus was obliterated

Boy, newborn (G J 521011) —The infant was transferred from the obstetric clinic on his first day of life because of hemolytic disease of the newborn. His respiration was extremely irregular and he was slightly cyanotic. During the first three weeks, a systolic murmur of varying intensity was heard over the pulmonary area. The auscultatory findings were subsequently normal. He was given repeated blood

[illegible]

tion of the respiration and an improvement in the general condition. At 15 days of age, cardiac catheterization was performed in association with an exchange transfusion. The catheter was introduced with the utmost caution into the pulmonary artery, it passed with ease through the ductus arteriosus into the descending aorta The relevant pressures and results of the gas analyses are shown in Table 13 The fact that the ductus was patent was demonstrated by the passage of the catheter The difference between the oxygen content of the right ventricle and that of the pulmonary artery was not sufficient to afford definite evidence of a shunt The most probable explanation of the murmur heard in the first three weeks of life is the patency of the ductus, the findings on cardiac catheterization give some support to this assumption

TABLE 13 [illegible]

Case	Pressure mm Hg			Vol %		Comments
	RV	PA	Aorta	RV	PA	
L.C. 520611 (4 weeks)	30/0	—	—	9 2	11 1	Air breathing
	30/3	30/18	—	10 0	10 4	100% O_2 breathing
G J 521011 (15 days)	33/0	37/22	70/60	9 8	10 4	Air breathing

The ductus subsequently closed spontaneously In case L.C 520611 the shunt decreased on breathing 100 per cent oxygen

Both the clinical observations and the results of cardiac catheterization indicate that asphyxia retards closure of a normal ductus arteriosus On the other hand, it is not known whether this condition is in any way associated with a persistent patent ductus In all of our cases, closure of the ductus took place eventually In connection with a follow-up study of the mental and physical development in 85 children with asphyxia neonatorum made by one of us in collaboration with Hellstrom, no case with a patent ductus was found (328)

ANATOMY —The ductus arteriosus varies greatly in width, length and shape It is

usual to differentiate a cylindric, a funnel-shaped, a window-shaped, and an aneurysmal ductus. In the funnel-shaped type, the ductus is wider at the aortic end and narrows toward the pulmonary artery. In the window-shaped and aneurysmal types, it forms a wide, direct communication between the aorta and the pulmonary artery. The ductus arteriosus runs from the aorta, *immediately below the origin of the left* subclavian artery, to the pulmonary artery,

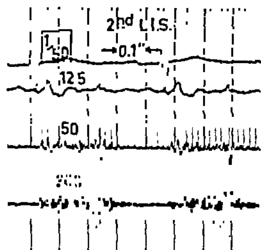


Fig. 479.—Patent ductus arteriosus. Phonocardiogram illustrating a typical continuous murmur in a 10-day-old girl (L C 520611). At 4 months of age, no murmur was audible Boxed figures denote degree of amplification, other figures denote standard frequencies of the filters L.I S, left interspace.

near the bifurcation. A right-sided ductus is extremely uncommon

Our series comprised 138 cases of patent ductus arteriosus 103 girls and 35 boys The preponderance of females in our series is in agreement with the general sex incidence of this cardiac malformation.

Cardiac catheterization was performed in only 47 of our cases, they therefore comprise selected material. They consisted partly of cases in which the clinical diagnosis was uncertain, or a coincident malformation was suspected, and partly of cases with marked clinical signs and symptoms. *Other cardiac anomalies were, in fact,* found in 12 of these cases: ventricular septal defect (two cases), ventricular septal defect and pulmonary stenosis (one case), atrial septal defect (one case), aortic stenosis (five cases), valvular pulmonary stenosis with stenosis of the right branch of the pulmonary artery (one case), infundibular stenosis with coarctation of the aorta (one case), and complete A-V block (one case). Thus, 126 cases of uncomplicated patent ductus arteriosus remain, the diagnosis was confirmed at operation in *every case. In addition,* angiocardiographic examination only was performed on four adults who were treated at another clinic and for this reason are not included in the clinical description.

Both age distribution and physical development are recorded in Figure 480 Those patients who showed the greatest degree of underdevelopment were also those with the most severe heart disease, with a large shunt, and with considerable enlargement of the heart.

CLINICAL FEATURES

Since the ductus arteriosus joins two arterial systems differing greatly in pressure *throughout the cardiac cycle*, a flow can take place during both systole and diastole. The resistance is occasionally greater in the pulmonary than in the systemic circulation, and the flow then goes from the pulmonary artery to the aorta. The blood may sometimes be shunted through the ductus only *during systole.*

The left to right shunt through the ductus implies an ineffective flow which places an additional burden on the left ventricle. This flow passes from the aorta *to the ductus* and continues through the pulmonary circulation to the left atrium and back to *the left ventricle.* These chambers and vessels therefore become dilated. If pressure in the pulmonary artery is high, the right ventricle is also overburdened and becomes hypertrophied.

The most common cause of death is heart failure, which does not usually appear until *the fourth decade* (133), but may occasionally appear as early as the first year of life (447). Many deaths have also been caused by bacterial endocarditis.

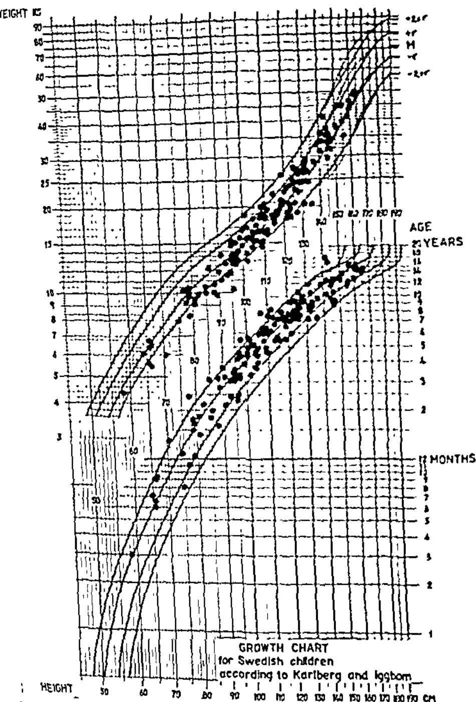


Fig 480 —Physical ... greatest ... of heart. ... Iggbom ...

The overwhelming majority of our patients were asymptomatic. Our series nevertheless comprises only children under 17 years of age, and symptoms seldom make their appearance before adult age. More or less marked symptoms were, however, present in 28 cases. The most common mani- [illegible] disability, and three of these patients were cyanotic. It is always exceedingly difficult to judge the statements in the history, and the normal physical tolerance of a child of a certain age is hard to determine. After an operation, the parents not uncommonly state that a marked improvement has taken place, even when, preoperatively, the child was said to "behave exactly like a healthy child." Consequently, it is probable that a larger number of children than the aforementioned actually had symptoms of some kind.

In our series, the age distribution was the same among those with and without symptoms. The symptoms had an early onset, in 10 cases during the first months of life. In certain cases the symptoms were first noticed after the child had started to walk, but in only two cases did an exacerbation occur during school age Of the 47 patients who underwent cardiac catheterization, 21 had symptoms and 26 were asymptomatic. Pulmonary hypertension was a characteristic feature in the patients with symptoms, all but three had a systolic pressure in the pulmonary artery in excess of 40 mm Hg, whereas pressure was raised in only four of the asymptomatic patients

The *physical working capacity* was determined in only nine cases It was low when pulmonary hypertension was present, but was normal in cases with normal or only slightly raised pulmonary artery pressure, even if the left to right shunt was large

Physical Signs

The most characteristic finding in patent ductus is the *continuous murmur* It was described for the first time by Gibson (276) in his classic lecture to medical students in Edinburgh in 1900. The murmur is both systolic and diastolic in time, it has a definite crescendo up to the second sound and then decreases in amplitude, waning in late diastole. It is usually localized to the second left interspace and is transmitted to the left axilla and to the interscapular region. The murmur is characterized by its accentuation during systole. In many cases there is a distinct pause after the end of the first sound, the murmur then begins in mid systole. On the phonocardiogram, this is manifested by a shift of the murmur to the right in the cardiac cycle. It was this shift, heard by Gibson on simple auscultation, that led him to infer that the murmur was on a vascular and not on a cardiac basis. In infants, the position of the murmur in the cardiac cycle can be determined only with difficulty on auscultation, owing to their rapid heart rate. It nevertheless appears distinctly on the phonocardiogram (167, 296, 462, 463, 466).

Figure 481 shows phonocardiograms in four typical cases of patent ductus arteriosus. *A* and *B* are from patients with a small patent ductus and a fairly insignificant shunt, whereas *C* and *D* are from patients with a wide patent ductus and a large shunt In the latter instance, the murmur is seen to fill the whole of the cardiac cycle. *It is probable that the* murmur occurs when the blood flows out of the patent ductus into the pulmonary artery. Flow through the ductus is greatest when the difference between the pressure of the aorta and the pulmonary artery is most marked, i e , when the pulse wave through the aorta has reached the ductus, about 0 02 sec after its start from the aortic orifice. This slight delay does not explain the fact that the maximum intensity of the murmur coincides with the second sound The flow is nevertheless retarded by a narrow ductus, and the maximal inflow to the pulmonary artery therefore occurs only with the second sound. When the ductus is extremely wide, the systolic accentuation occurs at the beginning of systole. A purely systolic murmur is sometimes heard in such cases, ow-

ing to the fact that a pressure gradient is present only during systole.

Cases in which the characteristic continuous murmur is lacking have sometimes been denoted as atypical, or the term atypical murmur has been used In our series, 15 such cases were found, cardiac catheterization disclosed considerable pulmonary hypertension in all of them The hemodynamic features of these cases are discussed in more detail later (p. 558) A mixed or a reversed shunt only was present in five of them, in two of these cases there was an extremely faint systolic murmur and in two a louder systolic murmur (Fig. 482). In one case there was a faint diastolic murmur only (Fig. 483). The other 10 patients, who had a left to right shunt only, presented a loud murmur which started immediately after the first sound and ended with the second sound In some of the cases there were, however, small vibrations at the beginning of diastole (Fig. 484). A murmur may be lacking entirely (603, 698).

A marked third sound or a short, low-frequency mesodiastolic murmur over the apex can often be heard and recorded in cases with a large left to right shunt

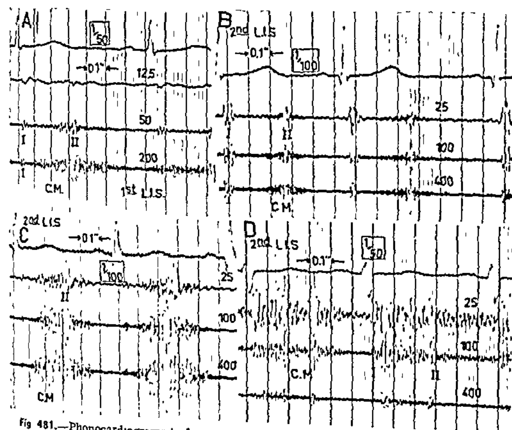


Fig 481.—Phonocardiograms in four typical cases of patent ductus arterios[illegible]
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LIS, left [illegible] CM, continuous murmur.

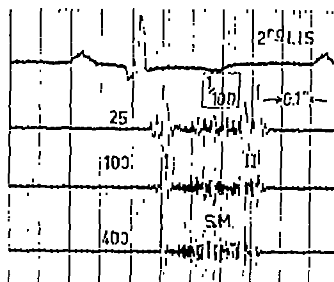
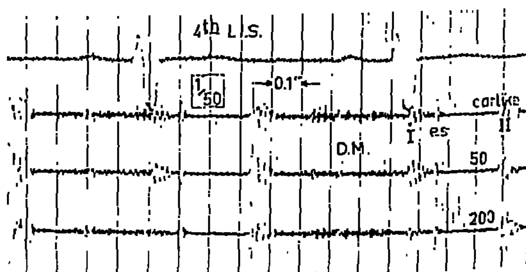


Fig. 482.—Phonocardiogram in atypical case of patent ductus arteriosus Girl, aged 7 (L.C. 460210), thin, with no cyanosis. Roentgenologic examination showed greatly enlarged heart Systolic murmur with slightly split, accentuated second sound, no diastolic murmur. Pressure. aortic 67/47 mm Hg, PA 82/54 (see p. 558). Mixed shunt. Oxygen saturation in the descending aorta, 84 per cent. Very large ductus, circumference at operation, 38 mm. Boxed figures denote degree of amplification, other figures denote standard frequencies of the filters. *II*, 2nd heart sound, *SM*, systolic murmur, *L I S*, left interspace.



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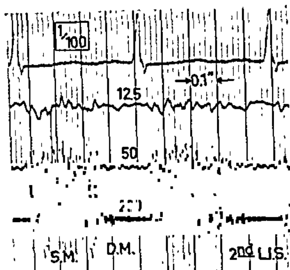


Fig 484.—Phonocardiogram in atypical case of patent ductus arteriosus. Boy, aged 3 (L.H 490418) Murmur begins with high amplitudes immediately after the 1st sound After the 2nd sound, there are only faint protodiastolic vibrations No continuous murmur Pressure. aorta 95/42 mm Hg, PA 80/50. At operation, the ductus was found to be as wide as the aorta. Left to right shunt After operation, a systolic murmur persists, probably due to valvular or subvalvular aortic stenosis Boxed figures denote degree of amplification, other figures denote standard frequencies of the filters *I*, 1st sound, *II*, 2nd sound, *DM*, diastolic murmur, *L I S* , left interspace, *SM*, systolic murmur

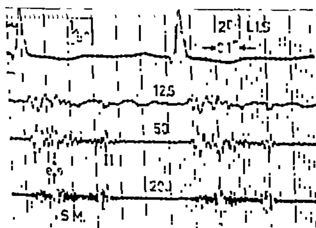


Fig 485 —Phonocardiogram ...
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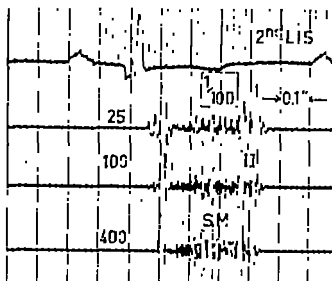
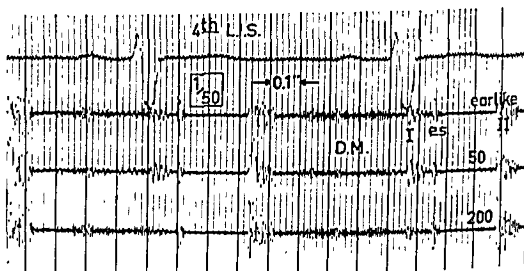


Fig. 482.—Phonocardiogram in atypical case of patent ductus arteriosus. Girl, aged 7 (L.C. 460210); thin, with no cyanosis. Roentgenologic examination showed greatly enlarged heart Systolic murmur with slightly split, accentuated second sound, no diastolic murmur. Pressure aortic 67/47 mm Hg, PA 82/54 (see p. 558). Mixed shunt Oxygen saturation in the descending aorta, 84 per cent. Very large ductus; circumference at operation, 38 mm. Boxed figures denote degree of amplification, other figures denote standard frequencies of the filters. *II*, 2nd heart sound, *SM*, systolic murmur, *L I S*, left interspace.



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is then usually heard over the whole thorax. (5) Venous hum of the neck. This murmur is easy to distinguish, since the diastolic component is strongest, it is heard most distinctly to the right of the sternum and above the clavicle, and better in the upright than in the recumbent position A venous hum on abnormal venous return to the left superior vena cava is, on the contrary, more difficult to differentiate from a patent ductus arteriosus (Fig 447, p. 491). (6) Multiple stenoses in the peripheral parts of the branches of the pulmonary artery (490) (7) A vascular communication between a coronary artery and the pulmonary artery (62).

A ventricular septal defect associated with aortic regurgitation may give rise to a murmur simulating that in patent ductus arteriosus (78, 292, 650, 705). Although we have no personal experience of such cases, we feel that it should be possible by phonocardiography to distinguish these murmurs from the typical or atypical murmur in patent ductus arteriosus

In the mildest cases of patent ductus, a faint continuous murmur is the only finding on clinical examination. If the murmur is louder, a thrill is also present With a large shunt, there is considerable enlargement of the heart Precordial bulge and a resistant, laterally displaced apex beat, of increased breadth, are then found Pulmonary hypertension causes right ventricular hypertrophy, the whole precordium is then heaving and the second sound over the pulmonary area accentuated. A large diastolic flow from the aorta to the pulmonary artery lowers the diastolic pressure in the systemic circulation, and the amplitude of the pulse becomes greater, as in aortic incompetence

Cyanosis is uncommon in patent ductus arteriosus Two of our patients had, however, exhibited cyanosis since the first months of life In these cases there was a reversed shunt, but no difference in the degree of cyanosis in the upper and lower half of the body could be observed In three other cases with a reversed shunt, no cyanosis was present

Electrocardiography

A narrow patent ductus with an insignificant shunt is associated with so small an [illegible]

When the shunt through the ductus is larger, signs of left ventricular hypertrophy arise If pulmonary hypertension is superadded, there is overburdening of the right ventricle as well, and signs of right ventricular hypertrophy also make their appearance

In our series isolated left ventricular hypertrophy was present in 21 cases, isolated right ventricular hypertrophy in five cases, and combined hypertrophy in 14. Cardiac catheterization was performed in 32 of the 43 cases with a pathologic electrocardiogram All of those patients with signs of left ventricular hypertrophy had a large left to right shunt, and all with signs of right ventricular hypertrophy had a systolic pressure in the right ventricle in excess of 50 mm Hg In one of the five patients with isolated right ventricular hypertrophy the condition was complicated by an atrial septal defect and in another by pulmonary stenosis, and in three cases there was high pulmonary vascular resistance The three last-mentioned patients also had abnormally high P waves in V_1.

The P-R interval was not prolonged in any of our cases, but in one case there was a congenital complete A-V block An incomplete right bundle-branch block was present in four cases

As in ventricular septal defect, the ECG picture varies considerably with the size of the shunt and the pulmonary artery pressure The ECG has no features specific to patent ductus arteriosus Consequently, it is of no great diagnostic value, although it gives a good idea of the degree of hemodynamic disturbances

Roentgenologic Examination

The increased blood flow in the aorta up to the mouth of the ductus arteriosus, in the

We found a systolic click (417) (see p. 419) in only six cases, but it may possibly have been masked in many cases by the murmur. It occurred 0.05 to 0.10 sec after the beginning of the first sound (Fig. 485).

A view generally held is that infants with a patent ductus arteriosus usually present a systolic murmur alone (177, 273, 557, 569, 650, 725). This is considered to be dependent on the low pressure gradient between the pulmonary artery and the aorta found at this age. In our experience, even In infants, the murmur varies with respiration and with the variation in intrathoracic pressure produced by it. On crying or forced breathing, the murmur may be inaudible or audible only in systole. It is often necessary to listen for some time, preferably when the patient is asleep as well, before the continuous murmur can be heard. As we have already mentioned, it may be difficult to hear the diastolic murmur in infants, owing to the rapid heart rate with shortening of diastole in particular. Consequently, the

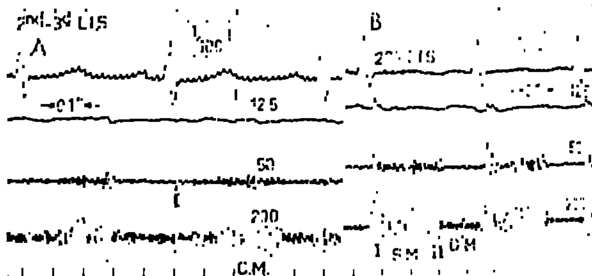


Fig 486.—Phonocardiograms in patent ductus arteriosus in infancy. *A*, boy, aged 6 months (R H 520302) No cardiac symptoms At routine examination, typical continuous murmur (*CM*) was heard Operation at 11 months of age showed a short, wide ductus with 30 mm circumfer- ... and retarded

with vibrations during diastole (*DM*), no typical con ... ter- gree of amplification, other figures denote standard ... space.

during the first month of life the pressure in the aorta is considerably higher than in the pulmonary artery during both systole and diastole, consequently, there is nothing to prevent a continuous murmur Our series includes nine children less than 12 months old, a continuous murmur was lacking in only three of them. The youngest patient, aged 3 months, had a typical continuous murmur. Three of the 10 children aged 1 to 2 years had an atypical murmur. These six patients with a systolic murmur only had a wide ductus and pulmonary hypertension. phonocardiogram often confirms the diagnosis in children of this age (Fig 486).

A continuous murmur can also be heard in other cardiac or vascular anomalies (296). The following are pertinent in the present connection: (1) Aortic pulmonary septal defect. (2) Aneurysm of a sinus of Valsalva with rupture into the right ventricle or atrium. (3) Arteriovenous fistula in the thorax. (4) Collateral circulation between the bronchial and the pulmonary arteries in tetralogy of Fallot with severe pulmonary stenosis or atresia. The murmur

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Electrocardiography

A narrow patent ductus with an insignificant shunt is associated with so small an extra burden on the heart that no signs of hypertrophy appear on the ECG. A normal ECG was found in 95 of our cases.

When the shunt through the ductus is larger, signs of left ventricular hypertrophy arise. If pulmonary hypertension is superadded, there is overburdening of the right ventricle as well, and signs of right ventricular hypertrophy also make their appearance.

In our series isolated left ventricular hypertrophy was present in 21 cases, isolated right ventricular hypertrophy in five cases, and combined hypertrophy in 14. Cardiac catheterization was performed in 32 of the 43 cases with a pathologic electrocardio-

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ROENTGENOLOGIC EXAMINATION

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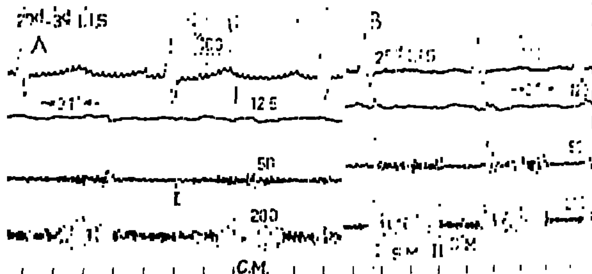


Fig. 486.—Phonocardiograms in patent ductus arteriosus in infancy *A*, boy, aged 6 months (*R H* 520302) No cardiac symptoms. At routine examination, typical continuous murmur (*CM*) was heard. Operation at 11 months of age showed a short, wide ductus with 30 mm circumference *B*, girl, aged 9 months (J W. 511218) Repeated, severe bronchopneumonia and retarded physical development High pressure in pulmonary artery (48/30 mm Hg) and aorta (103/37) At operation, the ductus was found to be even wider than the aorta Murmur is late systolic (*SM*) with vibrations during diastole (*DM*); no typical continuous murmur. Boxed figures denote degree of amplification, other figures denote standard frequencies of the filters. *L I S*, left inter space

A continuous murmur can also be heard in other cardiac or vascular anomalies (296). The following are pertinent in the present connection. (1) Aortic pulmonary septal defect. (2) Aneurysm of a sinus of Valsalva with rupture into the right ventricle or atrium. (3) Arteriovenous fistula in the thorax (4) Collateral circulation between the bronchial and the pulmonary arteries in tetralogy of Fallot with severe pulmonary stenosis or atresia. The murmur



Fig 487.—Patent ductus arteriosus Girl, aged 5 (A M 470903). Increased vascularity of the ungs, enlargement of left atrium and ventricle, no increased contiguity of surface of right ventricle to anterior wall of the thorax, indicative of hypertrophy, no apparent dilatation of aorta.

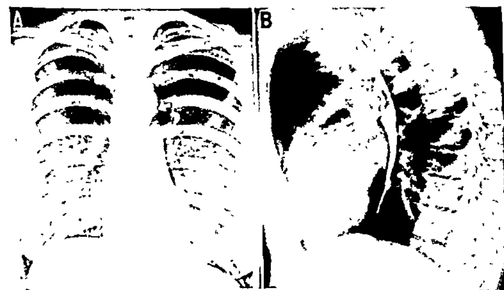


Fig 488 —Patent ductus arteriosus Woman, aged 26 (G P 270812), see Figure 508 (p 563). ncreased vascularity of lungs, enlargement of left atrium, slight dilatation of ascending aorta nd aortic arch, with distinct infundibulum (arrow in A)

pulmonary circulation, left atrium, and left ventricle causes dilatation of these vessels and chambers of the heart. Roentgenologic examination of 81 patients, 77 children and four adults, showed that the appearance is to some extent the same as in ventricular septal defect. However, in patent ductus arteriosus, the ascending aorta and the aortic arch are usually dilated and the right ventricle is not enlarged. In ventricular septal defect with a definite left to right shunt, *the aorta is of normal width or narrow and* the right ventricle is enlarged and hypertrophic. When patent ductus is complicated by pulmonary hypertension, hypertrophy of the right ventricle results. It is then nearly always difficult to make a roentgenologic differential diagnosis between ventricular septal defect and patent ductus.

UNCOMPLICATED PATENT DUCTUS ARTERIOSUS.—In the roentgenologic examination, two factors are of special importance: the width of the aorta and the size of its pulsations. It is not, however, invariably possible to identify the aorta on the roentgenograms in small children. It was clearly outlined in only 12 of our 25 patients under 3 years of age. During the first year of life, the aortic arch lies medially and may be *wholly or partly overlapped by the thymus;* consequently, it cannot be visualized without overlapping to the same extent as later. The aortic arch is occasionally distinct despite complete overlapping by the thymus. The evaluation is facilitated to some degree by contrast filling of the esophagus. Of 115 patients consisting of children over 3 years old and adults, 49 presented dilatation of the aorta, and in the rest it had a normal appearance (Figs. 487–491). The pulsations of the aorta were often distinctly increased, this could be observed in small children as well, although it was sometimes impossible to reach a definite decision, owing to their greatly increased heart rate and restlessness.

A conclusive finding is a local dilatation of the aortic arch (Figs. 488, 489, and 491). Its morphologic basis is a funnel-shaped widening of the aorta around the opening of the ductus arteriosus, denoted as the infundibulum (629). Jönsson and Saltzman (372) have pointed out that it can be demonstrated on ordinary roentgenologic examination and observed it in about 50 per cent of adult patients. It is not such a common finding in children. We observed it in only 20 of our 126 children. In the age group under 3 years, it was seen in one case only. It may be that the degree of dilatation of the infundibulum depends on the age of the patient, since it cannot be demonstrated *as frequently in older children as in adults.* In the youngest children, its visualization is also hampered by technical difficulties. Exposures are seldom obtained during maximal inspiration, and the aortic arch is therefore less prominent. A dilated infundibulum of the aorta is indicative but not pathognomonic of a patent ductus in children. We have observed in healthy individuals a structure whose appearance differed in no respect. Thus when a dilated infundibulum of the aorta appears as an isolated finding, which applied in four of our cases, it is of no decisive diagnostic importance.

Calcifications at the site of the ductus arteriosus were not seen in our series. They seem to have been described chiefly in older patients (474).

In most of our cases the main trunk of the pulmonary artery and its branches in the hilum were dilated, as an indication of an increased blood volume in the pulmonary circulation. The dilatation of the main trunk was seldom gross; it was usually moderate, not infrequently slight, and sometimes lacking entirely. The pulsations in the main trunk were distinctly increased in size in not a few of the cases, but they were only occasionally as conspicuous as in a large ventricular or atrial septal defect. The increased pulsations are presumably due partly to associated movements transmitted from the aorta. Another probable contributory factor is the blood flow from the ductus which, although it is continuous, varies in size during the cardiac cycle.

Left atrial enlargement is a characteristic feature of patent ductus arteriosus (201, 240). It is caused by the increased blood flow in the pulmonary circulation. The



Fig 487.—Patent ductus arteriosus. Girl, aged 5 (A M 470903). Increased vascularity of the lungs, enlargement of left atrium and ventricle, no increased contiguity of surface of right ventricle to anterior wall of the thorax, indicative of hypertrophy, no apparent dilatation of aorta.

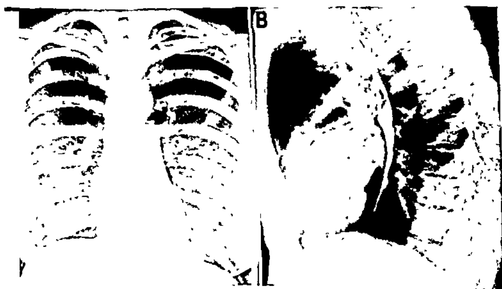


Fig 488 —Patent ductus arteriosus Woman, aged 26 (G P 270812), see Figure 508 (p 563). eased vascularity of lungs, enlargement of left atrium, slight dilatation of ascending aorta aortic arch, with distinct infundibulum (arrow in A).

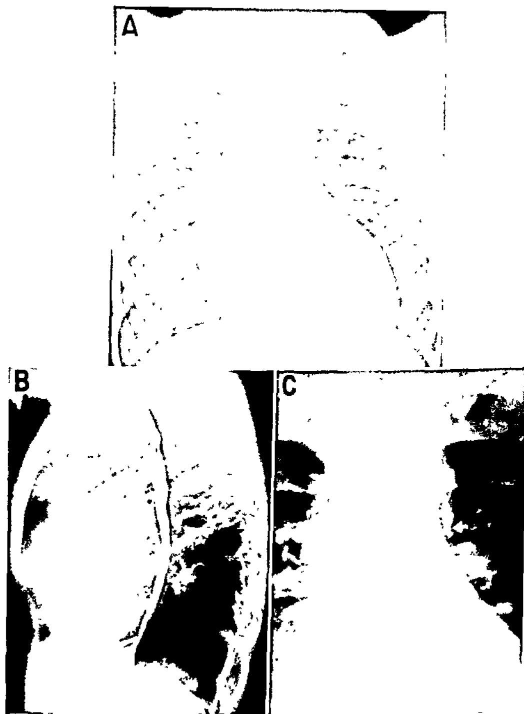


Fig. 489.—Patent ductus arteriosus with slight pulmonary hypertension Boy, aged 10 (G P 421217) Greatly increased vascularity of lungs, enlargement of left atrium and left and right ventricles, marked dilatation of ascending aorta and aortic arch, with distinct infundibulum (arrow in C)

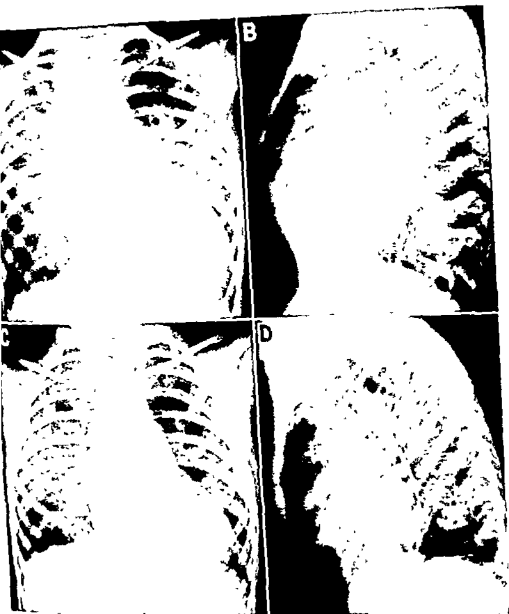


Fig 490 —Patent ductus arteriosus with very large left to right shunt Boy, aged 3 (L H 490418) A and B, before ligation of the ductus. Considerable increase in heart volume, with enlargement of left atrium and left and right ventricles, pronounced dilatation of pulmonary arter...

atrium was enlarged in 93 of our 104 cases with a left to right shunt only. No dilatation of the auricular appendage was found. When the shunt is small, the atrium is not enlarged. In 20 cases with typical clinical findings and a diagnosis verified at operation, the roentgenologic appearance was also normal in every other respect.

The enlargement of the left ventricle, which is due mainly to dilatation, may be difficult to judge objectively on roentgenologic examination. When the dilatation is distinct, there is increased curvature of the posterior and lateral border of the ventricle, and a depression of the apex into the diaphragm is often seen.

The increase in the heart volume is to be ascribed to the dilatation of both the left atrium and the left ventricle Gross enlargement was present in a few cases (Fig 490) In half of the cases the heart was definitely but moderately enlarged, and in the rest the values for the heart volume lay at the upper limit of the normal range of variations. Postoperatively, there was usually a rapid normalization of the heart volume.

The variations in the heart volume in a newborn infant with intermittent patent ductus arteriosus illustrate the effect of the shunt on the size of the heart.

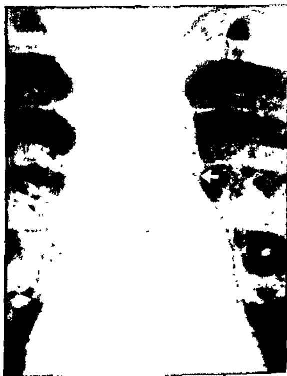
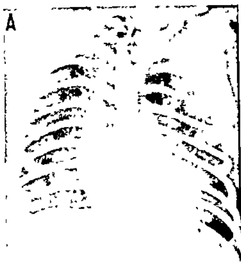


Fig. 491.—Patent ductus arteriosus. Girl, aged 14 (S S 401116). Considerable dilatation of ascending aorta and aortic arch, with distinct infundibulum (arrow).

[illegible] 492)

Two days later, a continuous [illegible] heard and the heart volume had increased to 54 ml. After a further seven days, the auscultatory findings were once more normal and the heart volume was 40 ml. At 9 months of age, both the heart volume and the physical findings were normal

Fig. 492.—Changes in roentgenologic appearance of heart in connection with opening and closure of the ductus arteriosus in a newborn child Girl (L C. 520611) A and B, on the 5th day of life, heart of normal size and outline, vascularity of lungs normal; auscultatory findings normal. C and D, on the 7th day, considerable increase in heart volume and vascularity of lungs, continuous murmur. E and F, on the 14th day, heart volume and vascularity of lungs approximately the same as on 5th day, decrease in size of left atrium, normal auscultatory findings. →





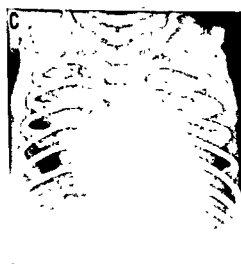








Fig 492 (legend on facing page)

PATENT DUCTUS ARTERIOSUS COMPLICATED BY SEVERE PULMONARY HYPERTENSION.—In 19 cases, pulmonary hypertension was recorded on cardiac catheterization. The roentgenologic appearance differed in certain respects in this group of patients (Figs. 493–495, 496, 497, and 498). There was increased contiguity of the heart surface to the wall of the thorax, sometimes with definite curvature of the anterior border and an upturned apex as indications of right ventricular enlargement and hypertrophy The pulmonary artery was, on the whole, more dilated. The cases with the most marked dilatation of the pulmonary artery were, in fact, found in this group Special investigations were required in several of the cases in order to make a differential diagnosis between ventricular septal defect and patent ductus with pulmonary hypertension When infundibular dilatation of the aortic arch could be demonstrated (Fig. 489) it was, however, of valuable diagnostic importance

In seven cases, the five children listed in Table 14 (see p. 558) and two adults, the shunt through the ductus was mixed or reversed. It is noteworthy that the ascending aorta and the aortic arch were not dilated in these cases (Figs. 494–497). In one case with a reversed shunt (Fig. 495), neither the left atrium nor the left ventricle was enlarged and the vascularity of the lungs was not increased. In the periphery of the lungs, the vessels were, in fact, narrow. In another case with a reversed shunt through the ductus and a left to right shunt through a ventricular septal defect (Fig 497), the appearance was the same as in a mixed shunt through the ductus (cf Figs 494 and 496). No calcifications were observed in the pulmonary vessels.



Fig. 493.—Patent ductus arteriosus with pulmonary hypertension. Girl, aged 10 (L R 430114), see Figure 575 (p 630) Moderate increase in heart volume and vascularity of lungs, enlargement of left atrium and left and right ventricles, curvature of apex, dilatation of pulmonary artery and aorta.

In these cases as well, the size of the left atrium varied consistently in relation to the blood volume in the pulmonary circulation The right atrium was enlarged in most cases.

Associated malformations were present in 12 cases They consisted of atrial septal defect, ventricular septal defect, ventricu-

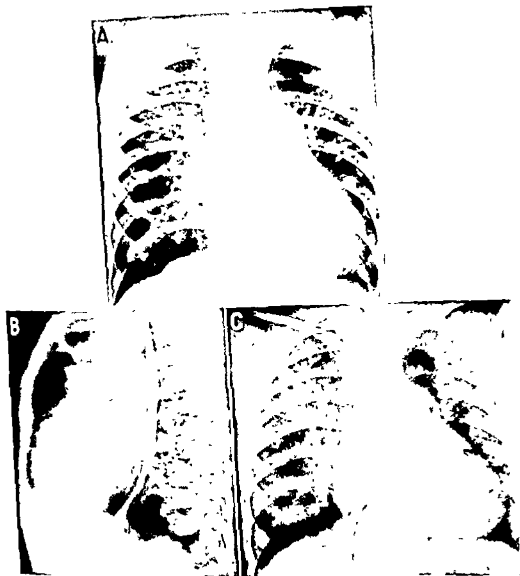


Fig 494.—Patent ductus arteriosus with equilibrated pressure in pulmonary artery and aorta Girl, aged 2 (I H 510424), see Figure 521 (p 574) Mixed shunt, moderate enlargement of left atrium, considerable enlargement of right ventricle great dilatation of pulmonary artery, none of the aorta. C, catheter passes through ductus into aortic arch and out into innominate artery.



Fig. 495.—Patent ductus arteriosus with equilibrated pressure in pulmonary artery and aorta Girl, aged 14 (R K 380309) Reversed shunt, no inc ... enlargement
ventricle,
enlargeme



Fig. 496 —Patent ductus arteriosus with equilibrated pressure in pulmonary artery and aorta. Boy, aged 6 (J A 471225), see Figure 519 (p 572) Mixed shunt, considerable enlargement of left atrium, right ventricle, and right atrium, greatly increased vascularity of lungs, no dilatation of aorta, infundibulum cannot be visualized

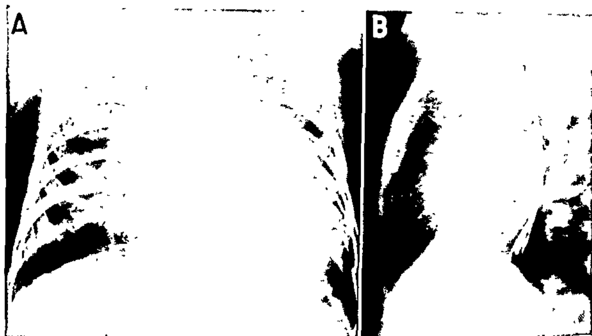


Fig. 497.—Patent ductus arteriosus with equilibrated pressure in pulmonary artery and aorta, and ventricular septal defect. Girl, aged 8 months (I N. 520422); see Figure 520 (p. 574). Reversed shunt through the ductus and a left to right interventricular shunt; great increase in heart volume with enlargement of left atrium in particular, greatly increased vascularity of lungs. Pulmonary artery is wide, but overlapping of the thymus makes it difficult to judge the width of both this artery and the aorta.



Fig. 498.—Wide patent ductus arteriosus and pulmonary hypertension. Girl, aged 11 (A B. 431028) The main trunk of the pulmonary artery (arrow) is considerably dilated Vessels in periphery of lungs narrow and somewhat irregularly delimited

lar septal defect and pulmonary stenosis (Fig. 499), valvular pulmonary stenosis (Fig 500), and aortic stenosis (two cases). They caused no specific changes in the roentgenologic appearance and were disclosed only on cardiac catheterization and angiocardiography.

ELECTROKYMOGRAPHY

A study of 26 cases showed that alterations were present chiefly in the electrokymograms of the aorta and the pulmonary artery.

In the group in which cardiac catheterization was not performed—in which any appreciable rise in the pulmonary artery pressure or a large shunt was presumably rare—aortic curves with a pathologic appearance were recorded in six cases and abnormal pulmonary artery curves in two. In the remaining 11 of the 17 cases studied in this group, the tracings were normal.

In the abnormal electrokymograms recorded over the ascending aorta, the characteristic features were absence of the peak in the upper part of the systolic upstroke and very slight prominence of the incisura and dicrotic wave (Fig. 501). They often lay far down on the descending limb. The onset of the systolic upstroke was sometimes early, and in some cases it was initially rounded, as in aortic incompetence. The most striking feature of the tracings of the aortic arch was the alterations in the descending limb (see Fig. 501).

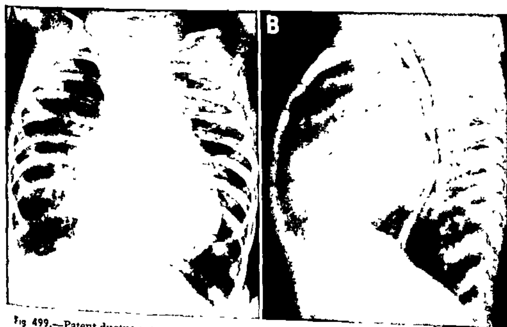


Fig. 499.—Patent ductus arteriosus ...

The pathologic pulmonary artery curves were characterized by a plateau in systole with superimposed deflections and by an irregularly shaped dicrotic wave (Fig 501).

In three cases with normal pulmonary artery pressure on catheterization, the electrokymograms were normal in the presence of a small shunt but abnormal when the shunt was large. In the pulmonary artery curves, a peak in early diastole was a typical finding (Fig 502). It is presumably a manifestation of the extra dilatation of the pul-



Fig. 497.—Patent ductus arteriosus with equilibrated pressure in pulmonary artery and aorta, and ventricular septal defect. Girl, aged 8 months (I.N 520422); see Figure 520 (p. 574). Reversed shunt through the ductus and a left to right interventricular shunt; great increase in heart volume with enlargement of left atrium in particular, greatly increased vascularity of lungs. Pulmonary artery is wide, but overlapping of the thymus makes it difficult to judge the *width of both this artery and the aorta.*

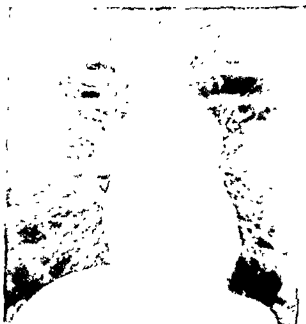


Fig. 498.—Wide patent ductus arteriosus and pulmonary hypertension. Girl, aged 11 (A B 431028). The main trunk of the pulmonary artery (arrow) is considerably dilated Vessels in periphery of lungs narrow and somewhat irregularly delimited

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ELECTROKYMOGRAPHY

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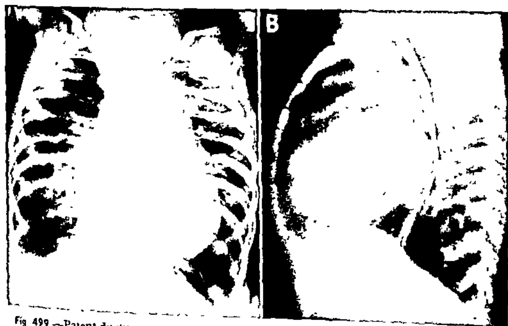


Fig 499 —Patent ductus arteriosus, valvular pulmonary stenosis, and defect in membranous part of the ventricular septum. Girl, aged 4 months (B K -S 521028), see Figure 372 (p 400). The roentgenologic appearance is dominated by a large left to right shunt Great dilatation of left atrium and enlargement of both ventricles Owing to the wide, overlapping pulmonary artery, width of the aorta cannot be evaluated

The pathologic pulmonary artery curves were characterized by a plateau in systole with superimposed deflections and by an irregularly shaped dicrotic wave (Fig 501).

In three cases with normal pulmonary artery pressure on catheterization, the electrokymograms were normal in the presence of a small shunt but abnormal when the shunt was large. In the pulmonary artery curves, a peak in early diastole was a typical finding (Fig 502) It is presumably a manifestation of the extra dilatation of the pul-

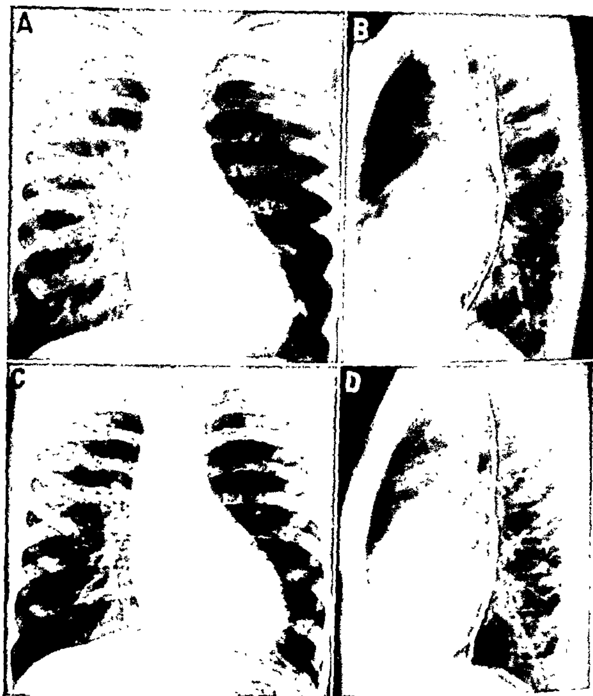


Fig. 500.—Patent ductus *arteriosus* and pulmonary stenosis Girl, aged 10 (M N 420410). *A* and *B*, before ligation of the *ductus* Roentgenologic appearance is entirely similar to that in patent ductus arteriosus with increased blood flow in the pulmonary circulation. No features are indicative of pulmonary *stenosis* *C* and *D*, after ligation of the ductus *Reduction* in heart volume and vascularity *of lungs* It is still *impossible* to identify features of pulmonary stenosis

onary artery, due to the blood flow rough the patent ductus.

With a left to right shunt through the uctus combined with pulmonary hyper-nsion, the electrokymogram of the aorta resented in five cases the same typical de-rmity as in the preceding groups (Figs. 03 and 505) In three of the cases, the

The electrokymogram from the left atrium in the same case showed a long contraction phase (0.10 sec), possibly ascribable to the large flow through the atrium.

Postoperative electrokymograms are illustrated in Figure 504.

In pulmonary hypertension with a right to left shunt through the ductus, the appear-

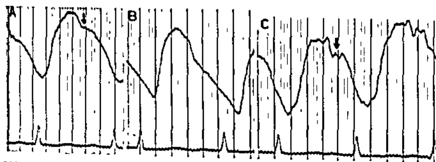


Fig 501.—Electrokymograms in patent ductus arteriosus
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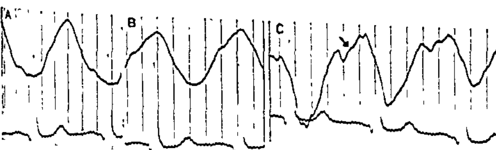


Fig 502 —Electrokymograms in patent ductus arteriosus Girl, aged 5 (A H 461118) A, ortic arch Incisura and dicrotic wave are absent B, left ventricle (apex) Short isometric ontraction phase Systolic deflection starts 0 05 sec after the Q wave C, pulmonary artery Posive extra wave in diastole (arrow) Normal PA pressure on catheterization

ppearance of the pulmonary artery curves as entirely or partly the same as in priary pulmonary hypertension Salient features were a prolonged systolic upstroke nd a reduced incisura and dicrotic wave ying high up on the descending limb A igh peak which in one case deformed the urve recorded over the vessels in the left ulum (Fig 503), was probably a manifestation of the shunt

ance of the electrokymograms in the single case investigated (Fig 506) differed from that in the others In the aortic tracings, the onset of ascent was late The upstroke ended, as *normally*, with a peak The incisura was distinct The dicrotic wave in the aortic arch *tracing* was both broader and higher than normally, possibly owing to the fact that the blood flow from the pulmonary artery is partly taken up in this segment of

the aorta. The electrokymogram of the pulmonary artery presented the same alterations as in pulmonary hypertension. The broad but low dicrotic wave and the markedly convex diastolic limb were particularly striking. Corresponding features were present in the tracing recorded over the left competence are recorded over the aorta.

In the pulmonary artery curves, a marked peak following the systolic upstroke (443) is probably a specific though inconstant feature. When pulmonary hypertension is present, typical pulmonary artery electrokymograms are often recorded with essen-

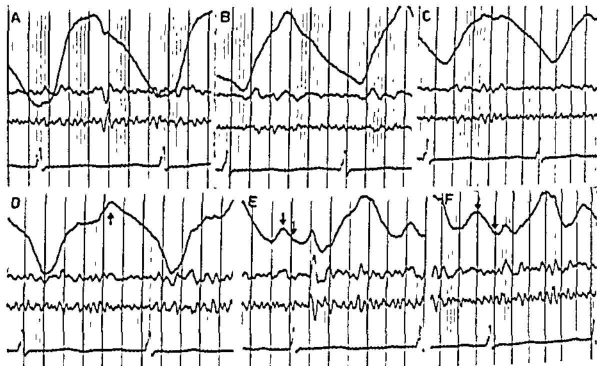


Fig. 503.—Electrokymograms in patent ductus arteriosus. Girl, aged 10 (L R. 430114) Frequency channels 10 and 20 cps *A*, ascending aorta *B*, aortic arch. In the aortic tracings, incisura and dicrotic wave are diminished Slow systolic upstroke in tracing from the aortic arch *C*, pulmonary artery Upstroke continues until the end of s lying directly below summit of the curve PA pressure *tolic peak (arrow) E, right atrium Tracing of norm* contraction, 0.06 sec, atrial contraction indicated by during contraction, 0.10 sec.

hilum. Electrokymographically, the right atrium showed no increased activity during presystole.

The abnormal electrokymograms of the aorta in patent ductus arteriosus with a left to right shunt are similar in several respects to those in aortic incompetence (213). They differ only with regard to the shape of the systolic rise. In patent ductus, its transit into the diastolic limb is curved, in aortic incompetence, it is usually terminated by a peak (*see* p. 663).

When there is a reversed shunt through the ductus, no tracings indicative of in- tially the same appearance as in other conditions with raised pressure in this artery

CARDIAC CATHETERIZATION

It is generally possible to diagnose a patent ductus arteriosus by means of clinical examination, phonocardiography, electrocardiography, and roentgenologic examination. The continuous murmur is the most salient feature Other cardiac malformations which cause a similar murmur are rare, and they seldom present the same clinical features. Moreover, the murmur

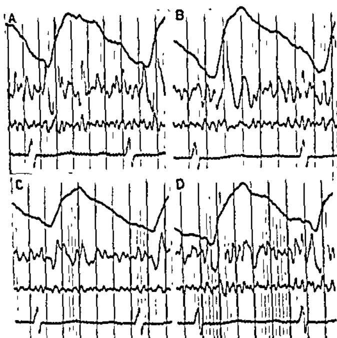


Fig 504 —Postoperative electrokymograms, same case as in Figure 503. *A*, ascending aorta Upstroke ends in a peak, incisura and dicrotic wave, which are still small, lie closer to summit of the curve than preoperatively *B*, aortic arch Upstroke has the normal steep slope, but incisura and dicrotic wave are still small *C*, pulmonary artery Incisura is at the normal site, dicrotic wave is small, upstroke has the normal gradient No postoperative catheterization. *D*, left hilum Tracing of almost normal appearance

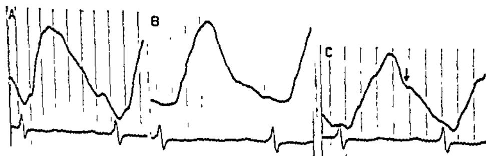


Fig 505.—Electr…
aged 9 (G P 421
pulmonary arter
wave PA pressu … mm Hg

differs somewhat in nature. It often has another localization. There is sometimes an accentuation during diastole, and the typical systolic crescendo around the second sound is frequently lacking. When patent ductus is combined with extremely high pressure in the pulmonary artery, and particularly when a reversed shunt is present, it is often impossible to diagnose it by the aforementioned methods. To establish the diagnosis in these atypical cases, cardiac catheterization and angiocardiographic ex-

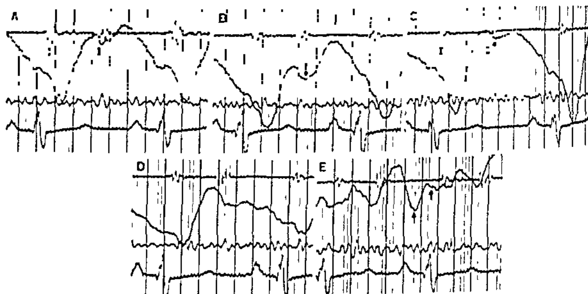


Fig. 506.—Electrokymograms in patent ductus arteriosus with reversed shunt. Girl, aged 16 ... *I* and *II*, 1st and 2nd pul- ... nd distinct incisura ... it of the 2nd pulmo- ... , synchronous with 2nd component of the 2nd pulmonary sound, dicrotic wave distorted and diastolic limb strongly convex *D*, left hilum Alterations of same nature as in *C* *E*, right atrium Ordinary curve, positive wave deformed by ventricular influence (between arrows) corresponds to atrial systole

amination are necessary The majority of our patients could be operated on without special investigations. But when the nature of the murmur or the other clinical features were in any way atypical, cardiac catheterization was performed. Even in cases in which the diagnosis was unquestionable *but we had reason to suspect* pulmonary hypertension, the patient underwent catheterization Thus, this examination was made in 47 of our cases.

The most definite evidence of a patent ductus is given by passage of the catheter through it. This was successful in 35 of our cases, and must mainly be ascribed to the fact that most of the patients who underwent this examination had an extremely wide ductus. However, it is in fact the cases with a wide patent ductus that are hard to diagnose clinically, and catheterization therefore offers a simple means of establishing the diagnosis. The position of the catheter must then be determined exactly, under fluoroscopic control in several projections. This seldom presents any difficulties The catheter passes from the pulmonary artery directly into the descending aorta. *In one case* it was, however, impossible to determine the position of the catheter with certainty The heart was rotated counterclockwise, so that the pulmonary artery lay to the right of the midline. *It was not until* angiocardiographic examination was performed that the catheter could be seen to have passed through a high ventricular septal defect into the ascending aorta

If the catheter does not penetrate the

ductus, it is necessary to rely on the gas analysis, which shows a higher oxygen content in samples from the pulmonary artery than from the right ventricle. The diagnostic value of this finding is nevertheless limited, for the following reasons. (1) The same condition applies in aortic septal defect (2) A similar result may be obtained in ventricular septal defect (see p 370). (3) When a patent ductus is combined with pulmonary incompetence, the oxygen content of samples from the right ventricle may be almost as high as that of samples from the pulmonary artery, but higher than that of samples from the right atrium Consequently, the condition may be interpreted as a ventricular septal defect It is therefore necessary to make an angiocardiographic examination to confirm the anatomic diagnosis

In pulmonary *hypertension*, it is of the utmost importance to determine the direction of the shunt through the patent ductus Samples must be *taken simultaneously* from the brachial artery and the descending aorta (or the femoral *artery*) When there is a reversed shunt, the oxygen content is higher in the vessels of the upper half of the body Burchell *et al* (122) have nevertheless demonstrated that the blood from the ductus can, to *some extent, also* pass into the aortic arch and into the upper half of the body

The pressure in the pulmonary artery gives a better conception of the degree of severity of the disease A marked correlation *is found* between the severity of the symptoms and the pulmonary artery pressure. With severe pulmonary hypertension, the picture changes, this applies to the clinical findings, the electrocardiogram, and the roentgenologic appearance

A narrow patent ductus is associated with such high local resistance and, consequently, with so large a fall in pressure from the aorta to the pulmonary artery that no pulmonary hypertension develops An extremely wide ductus presents such slight resistance in comparison with the peripheral resistance that there is a very small pressure gradient between the aorta and the pulmonary artery. The blood flow in the systemic and in the pulmonary circulation is then determined by the total resistance in the respective circulations. If the resistance in the pulmonary circulation were normal, the greater part of the output of the left ventricle would pass through the ductus into the pulmonary circulation, and no normal systemic flow could be maintained In cases with an extremely wide ductus, increased resistance in the pulmonary vascular bed is invariably present. The resistance may even be higher than in the systemic circulation, and the shunt through the ductus then becomes reversed. The shape and direction of the patent ductus are also of importance for the size of the shunt.

In 15 of our cases there was fairly severe pulmonary hypertension (pressure almost equilibrated with the systemic pressure during systole or diastole). The pulmonary artery pressure was probably not much increased in the 48 cases in which cardiac catheterization was not performed In eight cases the pressure in the pulmonary artery and the aorta was practically equilibrated during both systole and diastole. In two of these cases the shunt was reversed, and in three the shunt through the ductus was mixed A left to right shunt only was present in the remaining three cases, as well as in seven in which the diastolic pressure was *the same* in the pulmonary artery and the aorta, but the systolic pressure was higher in *the latter.* A continuous murmur was lacking in 12 of these 15 cases. Thus, a typical continuous murmur was audible in *three* cases despite equilibrated diastolic pressure In these cases there was a large left to right shunt When there is a *mixed* shunt the blood [illegible]
tion [illegible]
obser[illegible]
573 and 574) [illegible] pp.

Patent ductus arteriosus with marked pulmonary *hypertension* and a mixed or entirely reversed shunt is one of the forms of congenital heart disease that is difficult to diagnose *without* the help of cardiac catheterization In addition, it presents a

TABLE 14 — MAIN FINDINGS ON CARDIAC CATHETERIZATION IN 5 CASES OF PATENT DUCTUS ARTERIOSUS WITH A REVERSED OR MIXED SHUNT*

As a result of the low oxygen saturation in the lower half of the body the oxygen content of samples from the inferior vena cava is lower than that of samples from the superior vena cava

Case	O_2 Content, vol %							O_2 Cap., vol %	Pressure, mm Hg								
	SVC	IVC	RA	RV	PA	Desc Aorta	BrA		RA Mean	RV Syst	RV End-Diast	PA Syst	PA Diast	PA Mean	PCV Mean	Desc. Aorta Syst	Desc. Aorta Diast
Reversed shunt, R K 380309 (16 yr)	15.9	13.7	15.1	14.2	14.8	17.5	20.6	27.4	6	119	6	105	70	86	4	108	70
I N 520422† (8 mo)	9.9	9.4	9.3	12.7	12.9	13.4	—	16.5	0	75	2	75	37	58	10	75	30
Mixed shunt, L C 460210‡ (7 yr)	12.3	—	11.2	11.7	12.6	14.7	17.5	17.5	2	72	8	82	51	64	22	67	47
I H 510424 (22 mo)	9.9	8.9	9.1	8.8	10.0	12.4	13.2	15.9	3	66	5	67	41	56	15	72	29
J A 471225 (6½ yr)	9.9	8.1	9.1	8.8	12.4	13.6	14.3	15.0	6	66	9	66	40	55	14	80	40

*For abbreviations see Table 1, p. 119 †Combined with large ventricular septal defect ‡Combined with valvular aortic stenosis

great therapeutic problem. These conditions applied in five of our cases, and they will therefore be discussed in more detail The most important findings on catheterization may be inferred from Table 14. In every case the catheter passed through the patent ductus.

GIRL, AGED 16 (R K. 380309) —She had been cyanotic since birth. The cyanosis was equally marked in the upper and lower halves of the body. She was greatly disabled The shunt passed only from the pulmonary artery to the aorta. Oxygen saturation in the upper part of the body was also greatly decreased, this may have been due to oxygen unsaturation in the lungs or to passage of the blood from the ductus to the aortic arch

GIRL, AGED 8 MONTHS (I.N 520422).—Development was slow. She was dyspneic but not cyanotic. There was a large shunt from the left ventricle to the right, but no or only an insignificant shunt from the aorta to the pulmonary artery. No sample could be obtained from the brachial artery, but since the oxygen content of the descending aorta was low and the diastolic pressure in the pulmonary artery was higher than in the aorta, there was probably a right to left shunt through the ductus At operation, the ductus arteriosus was found to be as wide as the descending aorta The patient died 10 hours after ligation of the ductus. Autopsy disclosed a 7 mm wide defect in the muscular part of the ventricular septum

GIRL, AGED 7 YEARS (L.C 460210) —There was a shunt in both directions through the ductus At operation, a very slight coarctation was found distal to the ductus, it seemed to be of no functional consequence The ductus could be ligated without complications. At follow-up examination a systolic murmur was still present, but it had changed in nature, so that it was typical of aortic stenosis At exam-

but no hypertension was present in the upper part of the body Angiocardiographic examination showed valvular aortic stenosis and marked coarctation of the aorta *On direct* puncture of the left ventricle, a pressure of 224/28 mm Hg was recorded, with a systolic pressure gradient across the orifice of *100 mm* Hg The raised pressure in the pulmonary circulation remained on practically the same level as before operation, and was due *partly to the* increased filling pressure in the left ventricle.

GIRL, AGED 22 MONTHS (I H 510424) — Since her first year of life she had been cyanotic, to an equal degree in the upper and lower half of the body The shunt passed in both

directions through the ductus. At operation, an aneurysmal patent ductus was found. The patient died six days later. Pressure measurements during the operation showed the pulmonary artery pressure to be 75/46 mm Hg before closure of the ductus, and 40/24 after its closure

BOY, AGED 6½ YEARS (J.A. 471225) —He [illegible]

A continuous murmur was lacking in all of these five cases and no diagnosis could be made on the basis of the cardiac findings, the ECG or the roentgenologic appearance The picture was that of a large ventricular septal defect with pulmonary hypertension. The large blood pressure amplitude typical of patent ductus was also absent The diagnosis could be established by means of cardiac catheterization. A characteristic feature of these cases with a right to left or mixed shunt through the ductus was a lower oxygen content in the inferior vena cava than in the superior vena cava, contrary to the normal conditions

The resistance in the pulmonary vascular bed is increased There are anatomic changes *in the* small pulmonary arteries similar to those found in ventricular septal defect (p. 339) Judging by our material, high resistance *may* be present very early in life This is in agreement with the findings of other authors (11, 133, 175, 352, 612)

When resistance in the pulmonary circulation is so high that there is a right to left shunt only, the patent ductus has been regarded as a safety valve for the pressure in the pulmonary artery In such cases, the patients often die in connection with closure of the ductus (11, 175, 233, 287, 348, 612, 685) Although the flow from the pulmonary artery to the aorta through the patent ductus is altogether ineffective for oxygen transport, it is nevertheless of importance for maintaining the pressure in the systemic arteries In these patients, the vascular system has become adapted to a small pulmonary flow and a large systemic flow. After closure of the ductus, the systemic pressure must be maintained only by means of the flow through the pulmonary circulation. This flow must therefore be increased, and an increase in flow can occur only through an increase in pulse rate and a further rise in pressure in the right ventricle. The fall in arterial pressure and rise in pulse rate result in a poorer coronary flow, and the risk of ventricular fibrillation is great If the shunt through the ductus is mixed, particularly if the left to right shunt predominates, operation seems to be successful (87, 122, 352)

One of our two patients with only a right to left shunt was operated on (I.N. 520122). Her death shortly after operation cannot be ascribed to closure of a safety valve, because she still had a safety valve in the form of a large ventricular septal defect. One of our three patients with a mixed shunt died six days after operation, whereas the two others were operated on with considerable success A decisive question for the results of operation is whether the changes in the pulmonary vessels are reversible

At postoperative catheterization in three cases with considerable pulmonary hypertension, we found normal pressure in two cases and only slightly raised pressure in one The pressure had decreased from 100/60 to 26/8, from 65/30 to 20/5 and from 80/50 to 37/12 mm Hg, respectively. This decrease in pressure was more marked than could be ascribed to the decrease in pulmonary flow, indicating a decrease in resistance Similar results have been reported by many authors (12, 87, 122, 133, 228, 233, 601) A fairly long time, several months to a year, seems to be required for the pressure to become normalized Hultgren *et al* (352) reported a case in which the fall in pressure six weeks after operation was only 100/68 to 65/40 *mm Hg.*

Levinson et al (427) have described a characteristic pulmonary artery pulse pressure contour in patent ductus arteriosus,

TABLE 14.—MAIN FINDINGS ON CARDIAC CATHETERIZATION IN 5 CASES OF PATENT DUCTUS ARTERIOSUS WITH A *REVERSED* OR *MIXED SHUNT**

As a result of the low oxygen saturation in the lower half of the body the oxygen content of samples from the inferior vena cava is lower than that of samples from the superior vena cava.

Case	O_2 Content, vol %							O_2 Cap., vol %	RA Mean	Pressure, mm Hg							
										RV		PA			PCV Mean	Desc. Aorta	
	SVC	IVC	RA	RV	PA	Desc Aorta	BrA			Syst	End-Diast	Syst	Diast	Mean		Syst	Diast.
Reversed shunt, R K 380309 (16 yr)	15 9	13 7	15 1	14 2	14 8	17 5	20 6	27 4	6	119	6	105	70	86	4	108	70
I N 520422† (8 mo)	9 9	9 4	9 3	12 7	12 9	13 4	—	16.5	0	75	2	75	37	58	10	75	30
Mixed shunt, L C 460210‡ (7 yr)	12 3	—	11 2	11 7	12 6	14 7	17 5	17 5	2	72	8	82	54	64	22	67	47
I H 510424 (22 mo)	9 9	8 9	9 1	8 8	10 0	12 4	13 2	15 9	3	66	5	67	41	56	15	72	29
J A 471225 (6½ yr)	9 9	8 1	9 1	8 8	12.4	13 6	14 3	15 0	6	66	9	66	40	55	14	80	40

*For abbreviations see Table 1, p 119 †Combined with large ventricular septal defect. ‡Combined with valvular aortic stenosis.

great therapeutic problem. These conditions applied in five of our cases, and they will therefore be discussed in more detail. The most important findings on catheterization may be inferred from Table 14. In every case the catheter passed through the patent ductus.

GIRL, AGED 16 (R K. 380309) —She had been cyanotic since birth The cyanosis was equally marked in the upper and lower halves of the body. She was greatly disabled. The shunt passed only from the pulmonary artery to the aorta. Oxygen saturation in the upper part of the body was also greatly decreased, this may have been due to oxygen unsaturation in the lungs or to passage of the blood from the ductus to the aortic arch

GIRL, AGED 8 MONTHS (I N 520422) —Development was slow. She was dyspneic but not cyanotic There was a large shunt from the left ventricle to the right, but no or only an insignificant shunt from the aorta to the pulmonary artery No sample could be obtained from the brachial artery, but since the oxygen content of the descending aorta was low and the diastolic pressure in the pulmonary artery was higher than in the aorta, there was probably a right to left shunt through the ductus At operation, the ductus arteriosus was found to b as wide as the descending aorta The patien died 10 hours after ligation of the ductus Au topsy disclosed a 7 mm wide defect in the mus cular part of the ventricular septum.

GIRL, AGED 7 YEARS (L C. 460210) —Ther was a shunt in both directions through th ductus. At operation, a very slight coarctation was found distal to the ductus, it seemed to be of no functional consequence The ductus could be ligated without complications At follow-up examination a systolic murmur was still present, but it had changed in nature, so that it was typical of aortic stenosis At examination four years later, the symptoms of coarctation were accentuated. The arterial pulses in the lower part of the body were not palpable, but n
part c
tion
marked coarctation of the aorta On direct puncture of the left ventricle, a pressure of 224/28 mm Hg was recorded, with a systolic pressure gradient across the orifice of 100 mm Hg The raised pressure in the pulmonary circulation remained on practically the same level as before operation, and was due partly to the increased filling pressure in the left ventricle.

GIRL, AGED 22 MONTHS (I.H 510424) — Since her first year of life she had been cyanotic, to an equal degree in the upper and lower half of the body The shunt passed in both

directions through the ductus. At operation, an aneurysmal patent ductus was found. The patient died six days later. Pressure measurements during the operation showed the pulmonary artery pressure to be 75/46 mm Hg before closure of the ductus, and 40/24 after its closure

BOY, AGED 6½ YEARS (J.A. 471225) —He had never been cyanotic and was only slightly disabled There was a large left to right shunt through the ductus, and a small right to left shunt as well This mixed shunt could also be visualized on angiocardiography (Fig 519, p 573). At operation, an extremely wide ductus was found (1 5 cm in diameter). Operation was followed by marked improvement

A continuous murmur was lacking in all of these five cases and no diagnosis could be made on the basis of the cardiac findings, the ECG or the roentgenologic appearance The picture was that of a large ventricular septal defect with pulmonary hypertension The large blood pressure amplitude typical of patent ductus was also absent. The diagnosis could be established by means of cardiac catheterization. A characteristic feature of these cases with a right to left or mixed shunt through the ductus was a lower oxygen content in the inferior vena cava than in the superior vena cava, contrary to the normal conditions

The resistance in the pulmonary vascular bed is increased There are anatomic changes in the small pulmonary arteries similar to those found in ventricular septal defect (p. 339) Judging by our material, high resistance may be present very early in life. This is in agreement with the findings of other authors (11, 133, 175, 352, 612).

When resistance in the pulmonary circulation is so high that there is a right to left shunt only, the patent ductus has been regarded as a safety valve for the pressure in the pulmonary artery. In such cases, the patients often die in connection with closure of the ductus (11, 175, 233, 287, 348, 612, 635). Although the flow from the pulmonary artery to the aorta through the patent ductus is altogether ineffective for oxygen transport, it is nevertheless of importance for maintaining the pressure in the systemic arteries In these patients, the vascular system has become adapted to a small pulmonary flow and a large systemic flow. After closure of the ductus, the systemic pressure must be maintained only by means of the flow through the pulmonary circulation This flow must therefore be increased, and an increase in flow can occur only through an increase in pulse rate and a further rise in pressure in the right ventricle. The fall in arterial pressure and rise in pulse rate result in a poorer coronary flow, and the risk of ventricular fibrillation is great. If the shunt through the ductus is mixed, particularly if the left to right shunt predominates, operation seems to be successful (87, 122, 352).

One of our two patients with only a right to left shunt was operated on (I N. 520422). Her death shortly after operation cannot be ascribed to closure of a safety valve, because she still had a safety valve in the form of a large ventricular septal defect. One of our three patients with a mixed shunt died six days after operation, whereas the two others were operated on with considerable success. A decisive question for the results of operation is whether the changes in the pulmonary vessels are reversible.

At postoperative catheterization in three cases with considerable pulmonary hypertension, we found normal pressure in two cases and only slightly raised pressure in one The pressure had decreased from 100/60 to 26/8, from 65/30 to 20/5 and from 80/50 to 37/12 mm Hg, respectively. This decrease in pressure was more marked than could be ascribed to the decrease in pulmonary flow, indicating a decrease in resistance Similar results have been reported by many authors (12, 87, 122, 133, 228, 233, 601) A fairly long time, several months to a year, seems to be required for the pressure to become normalized. Hultgren et al (352) reported a case in which the fall in pressure six weeks after operation was only 100/68 to 65/40 mm Hg

Levinson et al (427) have described a characteristic pulmonary artery pulse pressure contour in patent ductus arteriosus.

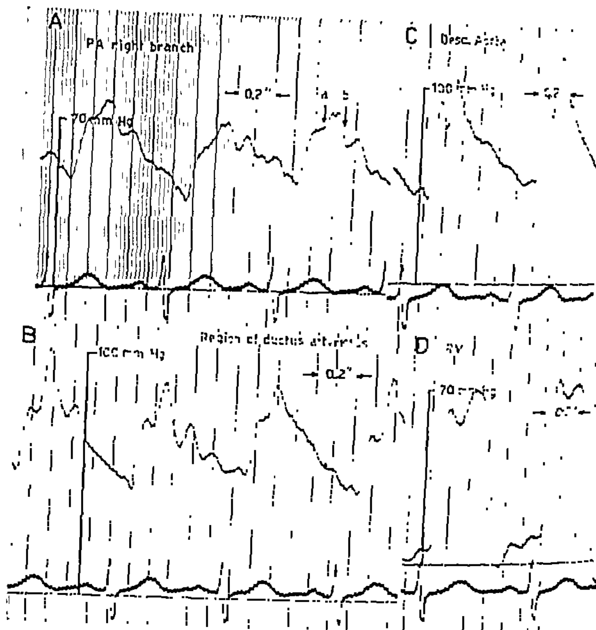


Fig. 507.—Patent ductus arteriosus Pressure recordings from right main branch of the pulmonary artery (*A*), region of the ductus arteriosus (*B*), descending aorta (*C*) and right ventricle (*D*) In *A* is seen an extra peak, it starts at *a*, ends at *b* and measures 7 mm Hg It is caused by the blood flow through the ductus from the aorta *B* was recorded after catheter was withdrawn from the right main branch of the pulmonary artery and was considered to lie in the main trunk The level and shape of the curve nevertheless agree with the curve from the descending aorta (*C*) On recording curve *B*, the catheter had thus slipped into the aorta When the ductus is short and wide, it may be impossible to determine on fluoroscopy whether the catheter tip lies in the aorta or the pulmonary artery

On continuous pressure recording when the catheter was withdrawn from the right branch of the pulmonary artery into the right ventricle, systolic and diastolic pressure rose on passage of the catheter past the region of the ductus arteriosus. The rise was considerable and was regarded as an effect of the transmission of systemic pressure through the patent ductus.

We also found the pulmonary artery pulse pressure curve to be characteristic in patent ductus. Figure 507 shows the pressure recording in a 4-year-old girl (K G 500327) with an extremely wide patent ductus. Marked respiratory variations can be noted. The curve from the right branch of the pulmonary artery (A) exhibits an extra systolic peak which starts 0 24 sec after the Q wave. This rise in pressure, which is only moderate (about 7 mm Hg), coincides with the summit of the pressure curve from the descending aorta (C) and therefore appears to be caused by the flow through the ductus. In relation to the velocity of the pulse wave from the left ventricle through the aorta and to the ductus, this extra peak on the pulmonary artery curve occurs later than the peak caused by the output of the right ventricle. Thus, an apparently unfeasible situation arises, in that the systolic pressure in the pulmonary artery is higher than in the right ventricle.

The pressure curve recorded when the catheter was withdrawn into the main trunk of the pulmonary artery at the mouth of the patent ductus (B) shows considerably higher pressure, both systolic and diastolic. This is strange, since the pressure gradient between the main trunk and a main branch cannot, as a rule, be measured. This rise in pressure in the main trunk cannot be ascribed to the fact that the tip of the catheter was directed against the flow from the patent ductus. A calculation of the pressure of velocity, based on the blood flow through the ductus and the width of the ductus shows that the rate of flow is so small that no measurable change in pressure can take place. The only plausible explanation of the high pressure in the main trunk is that the catheter tip did not, in fact, lie in the trunk, but that it had passed through the ductus, with the result that the pressure recorded was that of the aorta. The curve is in good agreement with the aortic curve (C). If the patent ductus is short and wide, the catheter can easily pass into the aorta when it is withdrawn from a branch of the pulmonary artery into the main trunk.

The large blood flow through the left side of the heart results in enlargement of the left atrium, and there is thus reason to anticipate a rise in pressure in this chamber. Consequently, it is of interest to record the PCV pressure. A satisfactory recording could be obtained in 28 cases, in 14 of them the mean pressure was between 10 and 20 mm Hg. In one case with an associated atrial septal defect and in two other cases with a patent foramen ovale, direct pressure measurements could be made in the left atrium. Patient R K., 380309, with a shunt only from the pulmonary artery to the aorta and consequently a small blood flow through the left side of the heart, had a low PCV pressure (4 mm Hg).

Pulmonary stenosis may sometimes be masked by a patent ductus, since the flow through the ductus raises the pressure in the pulmonary artery to the same level as that in the right ventricle. This rise in pressure occurs later in systole, with a maximum at the beginning of relaxation of the right ventricle. At the beginning of the ejection phase of the right ventricle, there is a pressure gradient between right ventricle and pulmonary artery, whereas the maximal systolic pressure level may be the same in both. Ekstrom (228) was able to demonstrate pulmonary stenosis in 12 cases in which a systolic murmur was present after operation for a patent ductus. Catheterization was not, however, performed preoperatively. In our series, pulmonary stenosis was diagnosed postoperatively in three cases, pressure recordings had been made before operation in two of them. One of the patients also had a ventricular septal defect, the pulmonary stenosis was not evident on catheterization until after the operation. In patient B K-S, 521028 valvu-

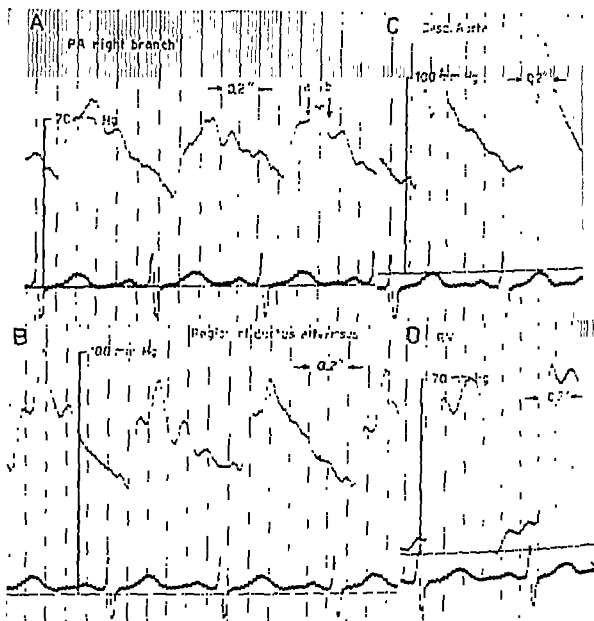


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lar pulmonary stenosis had, however, been *demonstrated on angiocardiography* prior to the operation (Fig. 372, p. 400). Table 15 shows the pressure in the right ventricle and pulmonary artery before and after operation, respectively.

In patient B.K-S. 521028, the pulmonary artery pressure did not become normalized after operation; this was due to the presence of the ventricular septal defect. Operation nevertheless resulted in a decrease in the total left to right shunt, and the condition became greatly improved. All of the

TABLE 15.—PATENT DUCTUS ARTERIOSUS AND PULMONARY STENOSIS: PRESSURE IN RIGHT VENTRICLE AND PULMONARY ARTERY BEFORE AND AFTER OPERATION

Case	Pressure, mm Hg			
	Before Op		After Op	
	RV	PA	RV	PA
M N 420410 (10½ yr.)	—	—	39/3	28/7
G.P. 421217 (9 yr.)	65/0	65/30	45/0	20/5
B K-S. 521028* (16 months)	74/0	68/21	71/8	50/8

*Combined with large ventricular septal defect

patients in question had only very slight pulmonary stenosis, of no great clinical importance.

In two other cases with pulmonary *stenosis demonstrated at angiocardiography*, catheterization was not performed after operation. In one case valvular stenosis had been present and the pressure gradient was only 7 mm Hg. In the other case the stenosis was infundibular, and the pressure gradient between the inflow tract of the right ventricle and the infundibulum was 22 mm Hg.

HEMODYNAMICS DURING EXERCISE

In cases with normal or only slightly raised pulmonary artery pressure at rest, a left to right shunt will take place during exercise as well. It can be inferred from Table 16 that the shunt does not necessarily increase as much as the systemic flow The

TABLE 16.—PATENT DUCTUS ARTERIOSUS: HEMODYNAMIC FINDINGS AT REST AND DURING EXERCISE IN ONE CASE*

Case No	Sex	Age, Year	Body Surface Area, m²	Physical Working Capacity, % of Predicted	Work Load, kg/min	O₂ Uptake, ml/min	Pulse Rate, beats/min	Pulmonary Circulation: AV O₂ Dif, ml/L	Pulmonary Circulation: Cardiac Output, L/min	Pulmonary Circulation: Stroke Vol, ml	Systemic Circulation: AV O₂ Dif, ml/L	Systemic Circulation: Cardiac Output, L/min	Systemic Circulation: Stroke Vol, ml	O₂ Cap, vol %	Art O₂ Sat, %	Shunt L/min L → R	RV Mean	Pressure, mm Hg: PA Syst.	PA Diast	PA Mean	Br A Syst.	Br A Diast	Br A Mean
69/56	F	34	1.55	79	Rest	260	88	16.1	16.2	184	34.9	7.5	85	17.9	96	8.7	33	26	10	16	150	68	91
					200	812	116	37.5	21.6	186	70.6	11.5	99	18.1	97	10.1	44	37	9	22	159	67	100
					400	1204	150	47.6	25.3	169	85.1	14.2	95	18.3	96	11.1	63	40	7	24	167	70	103

*For abbreviations see Table 1, p. 119.



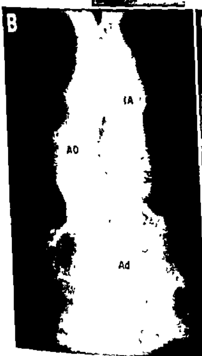




Fig 508 —Patent [illegible]

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ost [illegible]
aor [illegible]
[illegible] aortic infundibulum, PA, pulmonary artery,

lar pulmonary stenosis had, however, been demonstrated on angiocardiography prior *to the operation (Fig. 372, p. 400). Table 15* shows the pressure in the right ventricle and pulmonary artery before and after operation, respectively.

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69/56	F	34	1 55	79	Rest	260	88	16 1	16 2	184	34 9	7.5	85	17.9	96	8.7	33	26	10	16	150	68	91
					200	812	116	37 5	21.8	186	70 6	11.5	99	18.1	97	10.1	44	37	9	22	159	67	100
					400	1204	150	47 6	25.3	169	85.1	14 2	95	18 3	96	11.1	63	40	7	24	167	70	103

*For abbreviations see Table 1, p 110

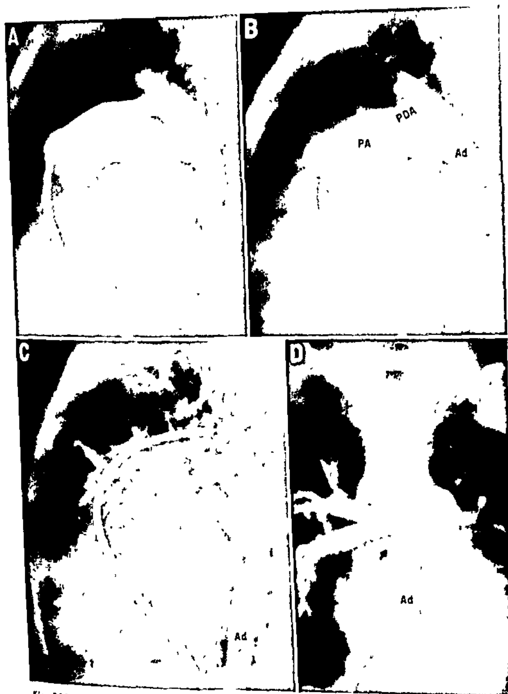


Fig 309 [illegible]

quantity of blood shunted per heart beat thus decreases during exercise. We have found this to apply in other cases in which the shunt was determined under a heavy work load. It is nevertheless likely that considerable individual variations occur, because in some cases the rise in pressure during exercise is more marked in the systemic than in the pulmonary circuit, with a resulting increase in the pressure gradient between aorta and pulmonary artery. No exercise tolerance test was made in any of our cases with high pulmonary vascular resistance, but Eliasch *et al.* (232) reported a case in which relatively heavy work was performed. Such work was found to be associated with a decrease in cardiac output in the pulmonary circulation.

ANGIOCARDIOGRAPHY

When the clinical findings have shown the presence of a typical patent ductus arteriosus, there are seldom indications for angiocardiographic examination An angiographic analysis of the vascular anatomy is not of such value from the surgical viewpoint that it outweighs the inevitable risks of the investigation. It is warranted in cases in which the clinical findings are questionable or when essential anatomic data are desirable When some associated malformation is suspected, the angiocardiographic examination should be planned with special consideration to this factor

Determination of the site and shape of the patent ductus presupposes its optimal filling with contrast medium Consequently, the examination should be made selectively, by injection of the contrast medium into the aorta when the direction of the shunt through the ductus is from left to right, and into the pulmonary artery when the shunt is reversed or mixed Overlapping by contrast medium-filled irrelevant structures is then reduced to a minimum, and this facilitates the interpretation still more

Angiocardiography was performed in 19 of our cases. Various methods were applied.

Thoracic aortography —This method, according to Jonsson *et al.* (369, 370), with injection of the contrast medium through a catheter with the tip placed in the ascending aorta, was used mainly in cases in which the catheter could not be passed through the patent ductus on right heart catheterization. Some typical findings are illustrated in Figure 508. This technique was used in seven investigations If the blood flow in the aorta is extremely large and the width of the radial artery does not permit the use of a catheter of sufficiently large caliber, the rate of injection will be too slow and the ductus cannot be visualized. The passage of contrast medium into the pulmonary artery at the site of the ductus nevertheless confirms the diagnosis

This method is presumably that most suited for disclosure of an aortic septal defect (298), but no such case was present in our material.

Thoracic aortography with modified technique (Jonsson *et al.* (369).—Contrast medium was injected through a catheter introduced into the ductus arteriosus via the pulmonary artery, with the tip placed in the aorta beside the mouth of the ductus (Figs 509 and 510). Passage of the catheter to the aorta at the site of the ductus is proof of patency of the ductus, but an analysis of the anatomy requires angiocardiographic examination

In order to obtain optimal filling of the ductus, we have in some cases withdrawn the catheter from the aorta into the pulmonary artery during the injection, which was therefore made partly into the ductus (Figs 511 and 512).

Injection of contrast medium into the left atrium —This was done through a catheter which had passed through an atrial septal defect or a patent foramen ovale The density in the aorta was not satisfactory with this technique, and it would be suitable only when the shunt is small

Injection of contrast medium into the right ventricle —This method was reserved for cases with complicating malformations, such as ventricular septal defect, infundibular stenosis, and valvular pulmonary stenosis. The prime object of the investigation was to identify these compli-



Fig 511.—*Patent* ductus arteriosus. Girl, aged 5 (S K. 480425) Injection of contrast medium during withdrawal of tip of catheter into the pulmonary artery (*A–D*). The small infundibulum is prolonged into a short, funnel-shaped ductus, about 3 mm in diameter (arrow in *D*). *Ad*, descending aorta, *AO*, aortic arch, *PA*, pulmonary artery

cating anomalies (Figs. 119 and 513). Valvular pulmonary stenosis is illustrated in Figure 514. During catheterization, no pressure gradient was recorded in the pulmonary orifice. The valvular stenosis was detected at angiocardiographic examination. The blood flow from the aorta causes dilution of the contrast medium in the pulmonary artery (281). When the shunt is small, a tiny filling defect is seen at the site aortic septal defect. The method provides no information about the anatomy of the ductus, and the dilution effect in the pulmonary artery should be regarded as a secondary finding. In none of the cases in which this technique was used was there sufficient density in the aorta, after the contrast medium had passed through the pulmonary circulation, to permit the identification of the patent ductus arteriosus,

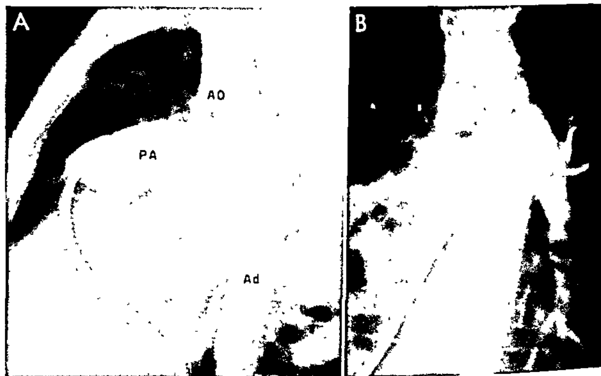


Fig. 510.—Patent ductus arteriosus Boy, aged 6 (Y M. 481005). The catheter tip lies in a short, wide ductus Strongly marked infundibulum (slightly below *AO*). *Ad*, descending aorta, *AO*, aortic arch, *PA*, pulmonary artery

of origin of the ductus (Fig. 186, p 194). With a larger shunt, there is diffuse dilution of the medium in the main trunk of the pulmonary artery (Fig 372, p 400), or a large blood deposit, containing little contrast medium, is visible (Fig. 515). The size of the filling defect varies greatly during different phases of the cardiac cycle, as stressed particularly by Wegelius and Lind (689). This phenomenon constitutes indirect proof of patency of the ductus, provided the filling defect appears at the typical site. When the shunt is large it is, however, impossible to rule out the possibility that the blood flow may be transmitted through an although reopacification only of the pulmonary artery supported the view that the shunt took place through the ductus

Elevation of the main trunk of the pulmonary artery and of its left main branch ... et al (629) ... o- ... id sus The finding is of an indirect ... is, according to our experience, difficult to evaluate objectively.

A partly patent ductus rarely seen is depicted in Figure 516, and an unusual anomaly of the right ventricle in a case of patent ductus arteriosus is demonstrated in Figure 517. The considerable dilatation of the left



Fig 514.—Patent ductus arteriosus and valvular pulmonary stenosis. Girl, aged 8 months (AJ 560127) Wide ductus with large left to right shunt. This produces great dilution of the contrast medium in the main trunk of the pulmonary artery (PA) The valvular pulmonary stenosis (halfway between *I* and *PA*) cannot be demonstrated on catheterization, owing to high pressure in pulmonary artery (41/18 mm Hg). *I*, infundibulum



Fig 515 —
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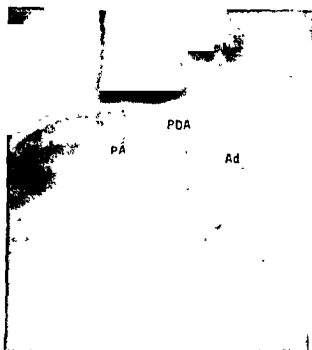


Fig. 512.—Patent ductus arteriosus. Boy, aged 10 (L K 431231). Injection of contrast medium during withdrawal of tip of catheter. Wide, short ductus, about 15 mm in diameter. *Ad* descending aorta, *PA*, pulmonary artery, *PDA*, patent ductus arteriosus

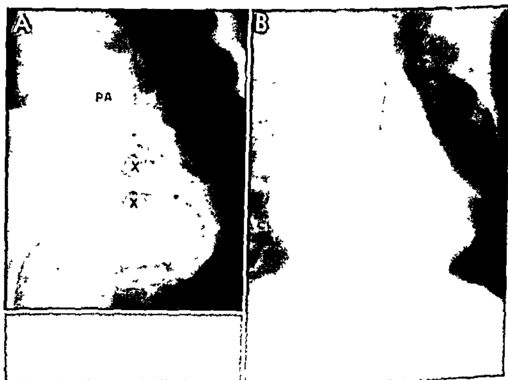


Fig. 513.—Patent ductus arteriosus and infundibular stenosis. Girl, aged 8 months (K B 550630). Constriction at the level of the ostium infundibuli (*X* in *A*) is probably caused both by displacement of the septal band and by adhesion of the trabeculae to the papillary muscles. On catheterization, the infundibular stenosis is masked by a wide patent ductus, through which a catheter is advanced into the descending aorta (*B*) *PA*, pulmonary artery

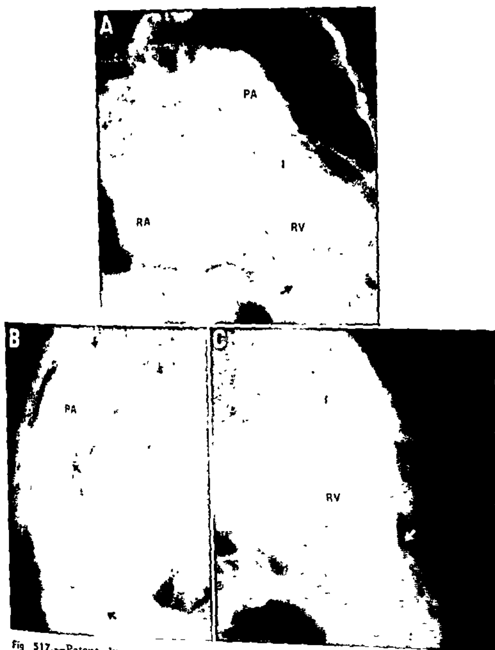


Fig 517.—Patent ductus arteriosus and anomaly of right ventricl... 460210) Intra... in B) in th... diverticul... B) At ope... gion ... artery



Fig. 516.—D[illegible] serted into left [illegible] 470225), with [illegible] aorta. *Ad*, desc[illegible] ascending aorta, *DA*, ductus arteriosus; *LSA*, left subclavian artery.

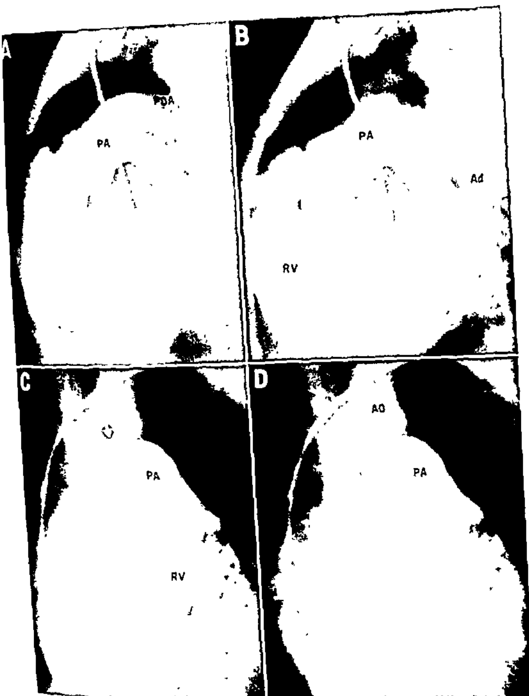


Fig 519b —Patent ductus arteriosus with equilibrated pressure in pulmonary artery and aorta Boy, aged 6 (I.A. 4719[illegible]) [illegible]
through the w[illegible]
thoracic aortog[illegible]
scending aorta[illegible]
RV, right vent[illegible]

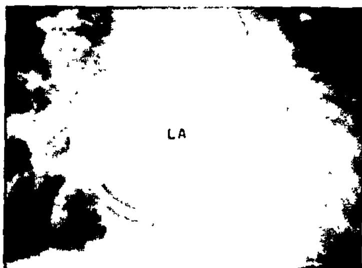


Fig. 518.—Patent ductus arteriosus Girl, aged 3 (K.Ö. 510303). Contrast medium injected into pulmonary artery. Owing to greatly increased flow in pulmonary circuit, both left atrium (*LA*) and pulmonary veins considerably dilated.

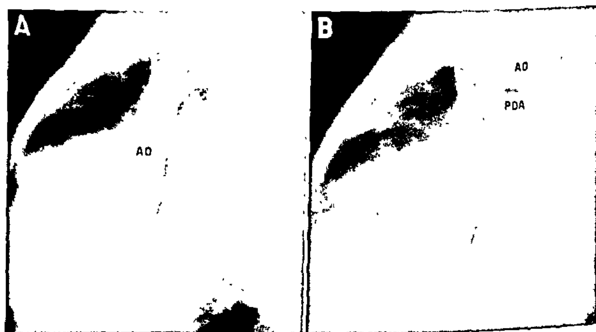


Fig 519a.—Same case as in Figure 519b Thoracic aortography *A*, diastole. Blood flow from the ductus arteriosus dilutes the contrast medium as far as into the ascending aorta *B*, systole Passage of medium through the wide ductus into the pulmonary artery *AO*, aorta; *PDA*, patent ductus arteriosus

atrium and pulmonary veins with a greatly increased flow is apparent in Figure 518

Injection of contrast medium into the pulmonary artery.—Such injection was made when the flow in the ductus was mixed or reversed Owing to the rise in pressure caused by the rapid injection, the ductus can be visualized even if the flow through it is large. This technique is illustrated in Figures 519 to 521. In Figure 519, during diastole, the shunt is directed toward the aorta, and both the aortic arch and the ascending aorta can be visualized in this phase During systole, the shunt changes its direction Aortography was also performed, the ductus could then be identified only during systole This observation is in agreement with the calculated resistances and the pressure curves from the aorta and the pulmonary artery.

It must, however, be borne in mind that the artificial conditions during angiocardiography do not permit any exact conclusions regarding the direction of the flow through the ductus in ordinary circumstances. The chief object of angiocardiography is visualization of the ductus, and conclusions relative to the circulatory physiology can be drawn only with great caution. This is illustrated by the findings in one of our cases with a reversed shunt through the ductus (Table 14 and Fig 520). During angiocardiography, which was performed under anesthesia and with the lungs saturated with oxygen, the shunt was mixed instead. It was directed during diastole toward the pulmonary artery and during systole toward the aorta. It is not improbable that the pulmonary resistance was altered by the high oxygen concentration, with a resulting change in direction of the blood flow during diastole.



Fig. 520.—Patent ductus arteriosus with equilibrated pressure in pulmonary artery and aorta Girl, aged 8 months (I N 520422). Injection into right ventricle *A*, diastole. Blood flow through the ductus from the aorta (arrow) *B*, systole Reversed flow, filling of entire descending aorta *Ad*, descending aorta, *PA*, pulmonary artery



Fig. 521.—Patent ductus arteriosus with equilibrated pressure in pulmonary artery and aorta Girl, aged 2 (I.H. 510424) Injection into right ventricle *A*, during diastole, right to left shunt [illegible] escending aorta *B*, widespread cystic with irregular distribution of the ves- [illegible] artery

atrium and pulmonary veins with a greatly increased flow is apparent in Figure 518.

INJECTION OF CONTRAST MEDIUM INTO THE PULMONARY ARTERY.—Such injection was made when the flow in the ductus was mixed or reversed. Owing to the rise in pressure caused by the rapid injection, the ductus can be visualized even if the flow through it is large. This technique is illustrated in Figures 519 to 521. In Figure 519, during diastole, the shunt is directed toward the aorta, and both the aortic arch and the ascending aorta can be visualized in this phase. During systole, the shunt changes its direction. Aortography was also performed, the ductus could then be identified only during systole. This observation is in agreement with the calculated resistances and the pressure curves from the aorta and the pulmonary artery

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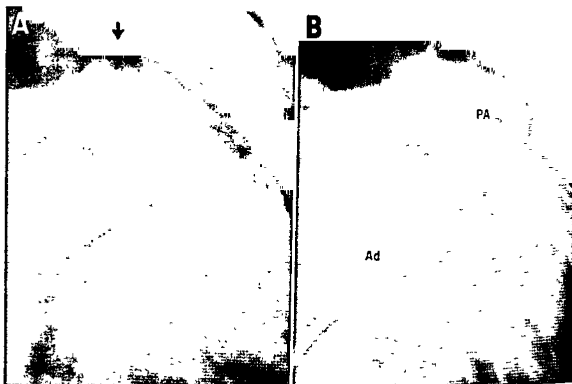
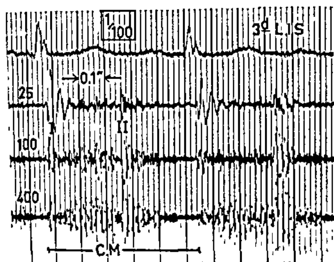


Fig. 520.—Patent ductus arteriosus with equilibrated pressure in pulmonary artery and aorta Girl, aged 8 months (I.N. 520422) Injection into right ventricle *A*, diastole. Blood flow through the ductus from the aorta (arrow). *B*, systole Reversed flow, filling of entire descending aorta *Ad*, descending aorta, *PA*, pulmonary artery.



F 521 ... artery and aorta ght to left shunt widespread cystic bution of the ves-



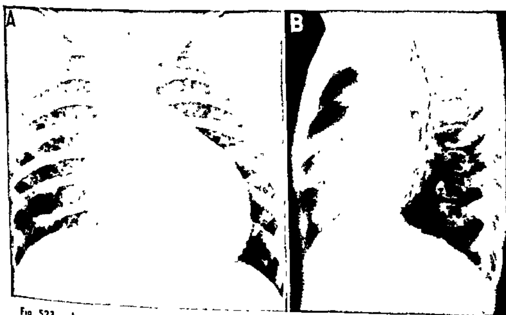


Fig 523.—Aortic sinus aneurysm with ... 430913), see Figure 525 M... left to right shunt, greatly ... aortic arch extremely narr...

17 Aortic Sinus Aneurysm with Communication to the Right Ventricle

ANEURYSM OF the aortic sinus may be congenital or acquired, in the latter case it is usually of syphilitic origin. In the congenital type, no symptoms are apparent until rupture takes place This is associated with the sudden development of a communication between the aorta and, as a rule, the right ventricle, but sometimes the right atrium or the pulmonary artery.

Both the anatomy and the clinical features have been described by Jones and Langley (362) in a survey of 25 cases of this malformation. The aneurysm was in the right aortic sinus in 20 cases and in the posterior sinus in five In 21 cases there was a communication with one of the chambers of the heart. No less than five were regarded as congenital, whereas rupture had occurred postnatally in 16 cases. When the aneurysm was in the right aortic sinus the communication was with the right ventricle, and when it was in the posterior sinus, with the right atrium.

According to Taussig (650), rupture takes place only after puberty, and the malformation would therefore never be manifest clinically in younger children. When rupture occurs, there is a sudden onset of severe symptoms and signs, which often lead rapidly to death. We have not observed any such case, but *our series* includes two cases of a congenital fistula between the aorta and the right ventricle.

GIRL, AGED 7 (M E. 450913).—A murmur had been present since birth She developed normally and was neither cyanotic nor disabled. The blood pressure was 110/50 mm Hg She had no precordial bulge or pathologic pulsations. The *cardiac findings* were completely dominated by an extremely loud, continuous murmur, which exhibited both a systolic and a diastolic accentuation and had a maximum over the third left interspace. The nature of the murmur thus differed somewhat from that in patent ductus. The phonocardiogram (Fig 522) showed that the systolic accentuation started immediately after the first sound, reached its maximum in midsystole, and waned toward the second sound. Immediately after the second sound, the murmur was once more accentuated and decreased slowly in intensity toward the middle of diastole (cf. the phonocardiogram in patent ductus arteriosus, p. 534). In so-called atypical patent ductus, the murmur has the same appearance during systole, but is not continuous, and the diastolic murmur is short and faint or is lacking altogether

The *electrocardiogram* showed slight left ventricular hypertrophy.

BOY, AGED 13 (E L 381031).—Heart disease was diagnosed at 1 year of age, but he had already exhibited cardiac symptoms. His physical development was retarded He was distinctly disabled, the chief complaint being dyspnea He had never been cyanotic. The blood pressure was 150/0 mm Hg. The *cardiac findings* were characterized by a pronounced precordial bulge and conspicuous pulsations over the precordium, as well as an *extremely* loud, continuous murmur with a maximum over the third to fourth left interspaces The phonocardiogram had the same appearance as in the previous case, except that the ampli-

both centrally and peripherally, were dilated The pulsations in the main trunk were conspicuous. The ascending aorta was markedly dilated, whereas the aortic arch was poorly visualized

CARDIAC CATHETERIZATION

The results of cardiac catheterization (Table 17) are the same as in ventricular (91 and 89 per cent, respectively). Such a decrease need not, however, be an indication of a right to left shunt, but may be due to oxygen unsaturation of the lungs, as we have observed in other cases with a left to right shunt (cf. Chapter 11, Ventricular Septal Defect).

Even if the results of cardiac catheterization seem to argue in favor of ventricular septal defect, this diagnosis cannot explain

TABLE 17.—AORTIC SINUS ANEURYSM WITH COMMUNICATION TO RIGHT VENTRICLE: RESULTS OF CARDIAC CATHETERIZATION IN 2 CASES

Case	O_2 Content, vol %						O_2 Cap	Pressure, mm Hg				
	SVC	IVC	RA	RV	PA	FA		RA	RV	PA	PCV	Aorta
M E. 450913 (7 yr)	11.2	12 8	11 4	12 8	12.8	15 0 = 91%	16 5	1	24/7	22/7	6	93/25
E.L. 381031 (13 yr)	12 9	—	11 2	14.7	14 0	89% sat	18 7	5	120/16	100/45	12	—

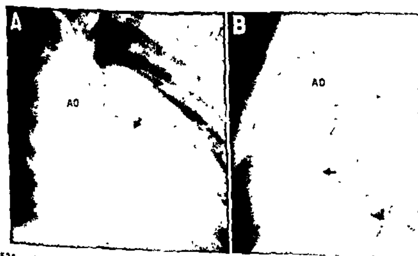


Fig 525.—Aortic sinus aneurysm with communication to right ventricle Girl, aged 7 (M E. 450913). Passage of contrast medium from left anterior sinus of Valsalva into right ventricle (arrows) AO, aorta

septal defect Patient M E., 450913, had a small shunt and normal pressure, this was in good agreement with the mild nature of the disease. Patient E L , 381031, who had severe symptoms, had a large shunt and elevated pressure *in the right* ventricle and pulmonary artery Arterial oxygen saturation was slightly decreased in both cases the murmur A patent ductus arteriosus with pulmonary incompetence or a ventricular septal defect with aortic incompetence can, on the other hand, present similar features Angiocardiographic examination is therefore required to establish the diagnosis On arterial catheterization for thoracic aortography it was possible in one case

tudes were considerably higher and the diastolic accentuation was more marked.

The *electrocardiogram* showed a complete right bundle-branch block, pronounced right ventricular hypertrophy, but also signs of left ventricular hypertrophy. The P waves were tall in V_{4R} and V_1.

A communication between the aortic sinus and right atrium via a dilated and tortuous branch of the right coronary artery was diagnosed by Grant *et al.* (298) at thoracic aortography. Cases with a communication between a branch of the left coronary artery and the right ventricle have been described by Baylis and Campbell (46) and by Davis *et al.* (180). A similar case with a communication to the pulmonary artery has been reported by Neill and Mounsey (504).

ROENTGENOLOGIC EXAMINATION

Little is known of the roentgenologic findings in aortic sinus aneurysm with a fistula. Expansile pulsations of the smaller branches (671) or of the main pulmonary arteries only (222, 109) have been described. In our two cases, the common features were increased vascularity of the lungs as a manifestation of a left to right shunt, as well as enlargement of the heart and a narrow aortic arch. The last-mentioned feature is in contrast to the conditions in patent ductus arteriosus

In the first case (Fig. 523), the enlargement of the heart was moderate It could be ascribed to the dilatation of both ventricles and the left atrium. The dilatation of the pulmonary artery was less conspicuous than could be expected to correspond to the increased vascularity of the lungs This was presumably due to the overlapping of the pulmonary artery by the dilated infundibulum of the right ventricle. The aorta was strikingly narrow

In the other case (Fig 524), the heart was grossly enlarged, owing to great dilatation of the ventricles and of the left atrium The pulmonary artery and its branches

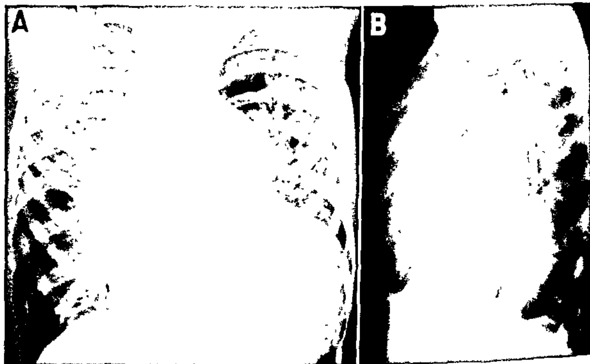


Fig. 524.—Aortic sinus aneurysm with communication to right ventricle. Boy, aged 13 (E L 381031), see Figure 526 Gross increase in heart volume, with enlargement of all chambers greatest of both ventricles, greatly increased vascularity of lungs, dilatation of main trunk of pulmonary artery. Ascending aorta is wide, aortic arch narrow.

both centrally and peripherally, were dilated The pulsations in the main trunk were conspicuous. The ascending aorta was markedly dilated, whereas the aortic arch was poorly visualized.

CARDIAC CATHETERIZATION

The results of cardiac catheterization (Table 17) are the same as in ventricular (91 and 89 per cent, respectively). Such a decrease need not, however, be an indication of a right to left shunt, but may be due to oxygen unsaturation of the lungs, as we have observed in other cases with a left to right shunt (cf. Chapter 11, Ventricular Septal Defect).

Even if the results of cardiac catheterization seem to argue in favor of ventricular septal defect, this diagnosis cannot explain

TABLE 17.—Aortic Sinus Aneurysm with Communication to Right Ventricle: Results of Cardiac Catheterization in 2 Cases

Case	O_2 Content, vol %						O_2 Cap	Pressure, mm Hg				
	SVC	IVC	RA	RV	PA	FA		RA	RV	PA	PCV	Aorta
M E. 450913 (7 yr)	11.2	12.8	11.4	12.8	12.8	15.0 = 91%	16.5	1	24/7	22/7	6	93/25
E L. 381031 (13 yr)	12.9	—	11.2	14.7	14.0	89% sat	18.7	5	120/16	100/45	12	—

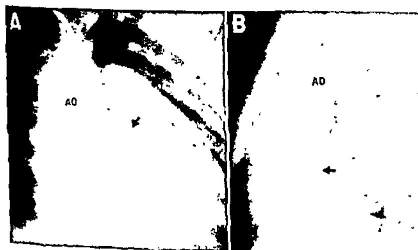


Fig 523.—Aortic sinus ane...
450913) Passage of contr...
(arrows) AO, aorta

septal defect Patient M E, 450913, had a small shunt and normal pressure, this was in good agreement with the mild nature of the disease Patient E L, 381031, who had severe symptoms, had a large shunt and elevated pressure in the right ventricle and pulmonary artery Arterial oxygen saturation was slightly decreased in both cases the murmur A patent ductus arteriosus with pulmonary incompetence or a ventricular septal defect with aortic incompetence can, on the other hand, present similar features. Angiocardiographic examination is therefore required to establish the diagnosis On arterial catheterization for thoracic aortography it was possible in one case

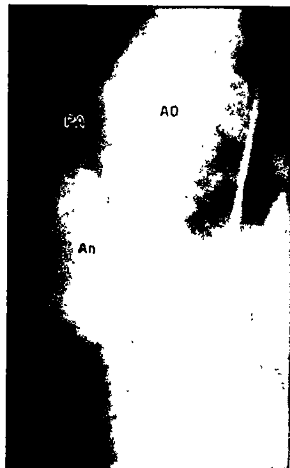


Fig. 526.—Aortic sinus aneurysm with communication to right ventricle. Boy, aged 13 (E L 381031) In front of and in contact with the aortic bulb is an egg-shaped cavity into which the contrast medium passed from the aorta. *AO*, aorta, *An*, aneurysm, *PA*, pulmonary artery. Diagnosis was confirmed at operation

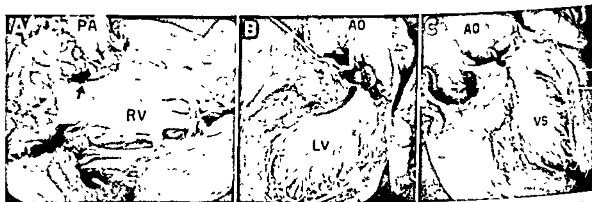


Fig. 527.—Aortic sinus a[illegible] ventricle Boy, aged 6 days (L L 440822). Wide commun[illegible] the aortic valve (arrow in C) [illegible] cle is dilated and thick-walled. *AO*, aorta, *LV* and *RV*, left and right ventricles, *PA*, pulm[illegible] artery, *VS*, ventricular septum.

ME 450913) to advance the catheter di-ectly from the bulb of the aorta into the ight ventricle, in which the pressure was roved not to be elevated.

ANGIOCARDIOGRAPHY

Thoracic angiocardiography in case M.E. 50913 (Fig 525) showed a leakage of ontrast medium from the left segment of he aortic bulb into the sinus part of the ight ventricle. The density was not, how-ver, sufficient to permit visualization of the istula The aortic sinuses were not de-ormed and the bulb of the aorta was of rdinary width, whereas the suprabulbar egment was narrow.

In patient E L, 381031, the contrast me-lium was injected into the right ventricle. No ventricular septal defect could be visu-alized, and, in view of the existing pressure conditions in the right ventricle, it should be possible to rule out such a defect on the basis of the angiocardiographic findings The pulmonary orifice was slightly widened and the pulmonary artery greatly dilated. The left atrium was enlarged. Concurrently with the visualization of the aorta, an egg-shaped aneurysmal formation was seen be-side the anterior segment of the aortic root (Fig 526); it bulged toward the lumen of the right ventricle below and to the right of the infundibulum. Recirculation of the contrast medium in the right ventricle could be demonstrated.

An autopsy specimen from a 6-day-old boy (L L, 440822), who was not included in our clinical series, is shown in Figure 527.

An aneurysmal sinus of Valsalva without a fistula to atrium or ventricle has been diagnosed by means of thoracic aortography (248) and venous angiocardiography (628). We have observed no such case.

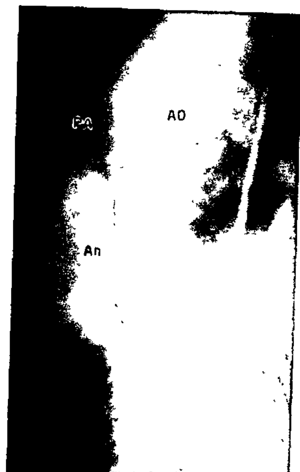


Fig 526.—Aortic sinus aneurysm with communication to right ventricle. Boy, aged 13 (E L 381031) In front of and in contact with the aortic bulb is an egg-shaped cavity into which the contrast medium passed from the aorta. *AO*, aorta, *An*, aneurysm, *PA*, pulmonary artery. Diagnosis was confirmed at operation.

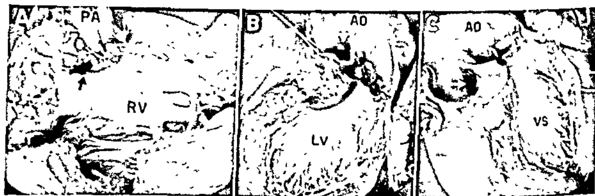


Fig. 527.—Aortic sinus aneurysm with communication to right ventricle Boy, aged 6 days (L.L. 440822). Wide communication (arrow in *B*), just over 5 mm, between left anterior cusp of the aortic valve (arrow in *C*) and infundibulum of the right ventricle (arrow in *A*). This ventricle is dilated and thick-walled *AO*, aorta, *LV* and *RV*, left and right ventricles, *PA*, pulmonary artery, *VS*, ventricular septum.

(M.E. 450913) to advance the catheter directly from the bulb of the aorta into the right ventricle, in which the pressure was proved not to be elevated.

ANGIOCARDIOGRAPHY

Thoracic angiocardiography in case M.E. 450913 (Fig. 525) showed a leakage of contrast medium from the left segment of the aortic bulb into the sinus part of the right ventricle. The density was not, however, sufficient to permit visualization of the fistula The aortic sinuses were not deformed and the bulb of the aorta was of ordinary width, whereas the suprabulbar segment was narrow.

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An autopsy specimen from a 6-day-old boy (L L, 440822), who was not included in our clinical series, is shown in Figure 527.

An aneurysmal sinus of Valsalva without a fistula to atrium or ventricle *has been* diagnosed by means of thoracic aortography (248) and venous angiocardiography (628). We have observed *no such case.*

18 Coarctation of the Aorta

IT IS usual to distinguish between an "infantile" and an "adult" type of coarctation of the aorta. These terms (*type ordinaire du nouveau-né* and *type ordinaire de l'adulte*), introduced by Bonnet (79) in 1903, were based on the anatomic conditions. According to Bonnet, the infantile type is characterized by an elongated constriction between the left subclavian artery and a patent ductus arteriosus, and is often combined with intracardiac malformations. The adult type is stated to present only a short constriction immediately below the origin of the left subclavian artery. Taussig (650) points out that "each of these types is a distinct anatomical entity" and that "the two conditions present entirely different clinical syndromes"

This classification has been subject to some criticism. The original anatomic distinction cannot be regarded as adequate. The "infantile" type is also encountered in adults, and the "adult" type in the newborn Edwards (216) has suggested the following classification instead:

1 Coarctation of the aorta with closed ductus arteriosus
   a) Coarctation in the vicinity of the ligamentum arteriosum
   b) Coarctation in unusual locations
   c) Coarctation with stenosis of the right or the left subclavian artery
2. Coarctation of the aorta with patent ductus arteriosus
   a) Coarctation distal to the aortic mouth of the ductus
   b) Coarctation proximal to the aortic mouth of the ductus
      (I) Without systemic collaterals
      (II) With systemic collaterals

This classification, based on the anatomic findings, is also adequate as far as the clinical syndrome is concerned, since the development of the collateral circulation and the position and width of the ductus arteriosus play a prominent role in the hemodynamics. Coarctation of the aorta is not uncommonly associated with a patent ductus, according to Abbott (1), this applies to 9 per cent of all cases. The ductus is often so narrow that no shunt can be demonstrated on cardiac catheterization, even if the mouth of the ductus is proximal to the coarctation, with a resulting large pressure gradient. The direction of the shunt through a ductus situated distal to the coarctation is determined by the development of the collateral circulation. If it is extensive, the shunt may go from the aorta to the pulmonary artery, but if collaterals are lacking and the coarctation is severe, the blood is shunted from the pulmonary artery to the descending aorta.

Bahn *et al* (33) have stressed the importance of the collateral circulation. They stated that in cases with a ductus arteriosus proximal to the coarctation, collaterals are formed *in utero*, but when the ductus lies distal to the coarctation, the fetal circulation is not affected and therefore no collaterals are developed They described three

ses belonging to the latter group; death occurred at the age of 3, 7, and 25 months, respectively. In all three cases, the collaterals were poorly developed The authors considered the cause of death to be left ventricular failure due to obstructive hypertension.

It is nevertheless probable that collaterals can develop *in utero* even when the ductus is distal to the coarctation If coarctation is severe and the ductus of the usual width, the blood flow through the ductus is insufficient to supply the descending aorta, including the placental circulation. If the ductus is abnormally wide, i e , malformed, the fetal circulation can, on the contrary, be maintained without collaterals Similarly, no collaterals are developed if the coarctation is only of moderate degree The type of malformation which allows the fetal circulation to be maintained without the aid of collaterals is associated with a poorer prognosis after birth, whereas the type in which collaterals are formed *in utero* is likely to function better postnatally

The clinical syndrome varies according to the degree of coarctation and the width of the ductus An extreme case is a malformation consisting of complete interruption of the aortic arch in combination with an abnormally wide ductus and no collateral circulation The fetal type of circulation through the ductus then persists postnatally From the clinical viewpoint, it is essential to distinguish between cases with a fetal direction of the circulation and those with a left to right shunt through the patent ductus The former present entirely divergent features, whereas the latter do not differ to any great extent from cases with a closed ductus A left to right shunt may occur even when the ductus is distal to the coarctation Such cases have been described by Taylor *et al* (653), Johnson *et al* (292) and Gøtzsche (359) One of us had the opportunity, at another hospital, of performing cardiac catheterization in a case of this kind, which is not included in our series The systolic pressure in the pulmonary artery was 35 mm Hg The catheter passed through the patent ductus into the descending aorta, in which the systolic pressure was 60 mm Hg. There was a moderately large left to right shunt through the ductus.

In the following discussion, coarctation of the aorta will be classified into two main groups:

A. Coarctation of the aorta; the ductus may be closed or patent. When it is patent, the shunt goes from the aorta to the pulmonary artery.

B. Coarctation of the aorta with a fetal type of circulation through the patent ductus

A COARCTATION OF THE AORTA

The term "adult type" of coarctation of the aorta is still more misleading than the term "infantile type." Apart from the fact that the "adult" type may also be found in the newborn, the term is apt to be associated with the so-called skodiac hypothesis concerning the pathogenesis of coarctation. According to this hypothesis, the constriction arises in connection with closure of the ductus arteriosus The smooth muscles which cause this closure are said to involve the aortic wall as well. This theory is not, however, tenable (216). As in other forms of congenital anomalies, it is presumably a matter of an embryonic malformation, consequently, it is incorrect to speak of an "adult" type.

Other cardiac malformations are often found in combination with coarctation of the aorta The most common is a bicuspid aortic valve. Abbott (1), for example, in her series of 183 cases found malformations of the aortic cusps of a definitely congenital type in 25 1 per cent (46 cases) Reifenstein (551) gave a still higher incidence, i e , 42 3 per cent A bicuspid aortic valve does not in itself necessarily result in any functional disturbance, but incompetence of the cusps is very likely to occur on the basis of the hypertension and dilatation of the aorta caused by the coarctation Obviously, this is aggravated by bacterial endocarditis, which frequently occurs in such patients Reifenstein (551) found a bi-

cuspid aortic valve in almost three-quarters of his cases of bacterial endocarditis. Aortic incompetence gradually appeared as a result of sclerosis of the bicuspid valve in no less than one-fourth of the cases in a large series studied by Campbell and Baylis (134). Aortic stenosis may also develop upon congenital bicuspid valves (611). As regards other complicating malformations, Abbott (1) found a patent ductus in 9 per cent, an atrial septal defect in 3 per cent, and subaortic stenosis in 2 per cent. According to Wood (713), rheumatic mitral stenosis is not uncommon.

The anatomy of the coarctation varies greatly, this applies to its localization and extent and to the degree of constriction. From the clinical and hemodynamic aspects, the degree of stenosis and extent of the collateral circulation are the features of main interest. From the surgical viewpoint, on the contrary, the anatomic conditions play the most important role. They will be discussed with the description of the angiocardiographic findings

In coarctation of the aorta, the symptoms can be referred to (1) hypertension in the upper part of the body (often cerebral symptoms), (2) the low pulse pressure in the lower part of the body, (3) cardiac symptoms caused by the increased burden on the left ventricle Many authors have stressed that the symptoms are late in appearing, usually only in adulthood (85, 423, 650), whereas others have found marked symptoms at an early age, sometimes even in infancy (33, 118, 129, 306, 383, 388, 407, 500, 565).

According to Abbott (1) and Reifenstein (551), the mortality rate is low during the first 10 years of life and rises during the second decade. During the first decade, death is usually due to bacterial endocarditis or complicating anomalies (e g , subaortic stenosis), but during the second decade, rupture of the aorta and intracranial hemorrhages become increasingly frequent. It is not until middle age that heart failure is one of the main causes of death

There are 61 cases of coarctation of the aorta in our series. In addition, 34 adults from other clinics underwent roentgenologic examination alone; only the roentgenologic features of these cases will be discussed. In five of our 61 cases a complicating congenital malformation was present, i.e., mitral incompetence (one case), aortic incompetence (one case), mitral stenosis (one case), aortic and mitral incompetence (one case), ventricular septal defect and endocardial fibroelastosis (one case). At operation the ductus arteriosus was found to be patent in 22 cases, but in most of them the lumen was very narrow. A left to right shunt was established on preoperative cardiac catheterization in only 17 cases. In every case the ductus took its origin from the aorta, proximal to the coarctation. In only three cases was the difference between the oxygen content in the right ventricle and in the pulmonary artery more than 1 volume per cent. A continuous murmur was present in 10 cases

Our series consisted of 38 boys and 23 girls, a preponderance of males has also been found in other series.

CLINICAL FEATURES

Taussig (650) has pointed out that children with coarctation of the aorta are strikingly heavily built and often have a healthy appearance The physical development in our series is recorded in Figure 528. Only two patients were overweight. Height was normal There was no underdevelopment of the lower part of the body.

Only 14 children had symptoms. Slight disability was present in three Five patients suffered from headache; they all had marked hypertension. Heart failure was present in three patients with associated malformations one with a large left to right shunt through a patent ductus, combined with pulmonary hypertension; one with aortic and mitral regurgitation, and one with ventricular septal defect and endocardial fibroelastosis The last-mentioned patient died at the age of 6 weeks. An additional two patients were severely disabled and suffered from dyspnea. One of them had mitral incompetence and the other a

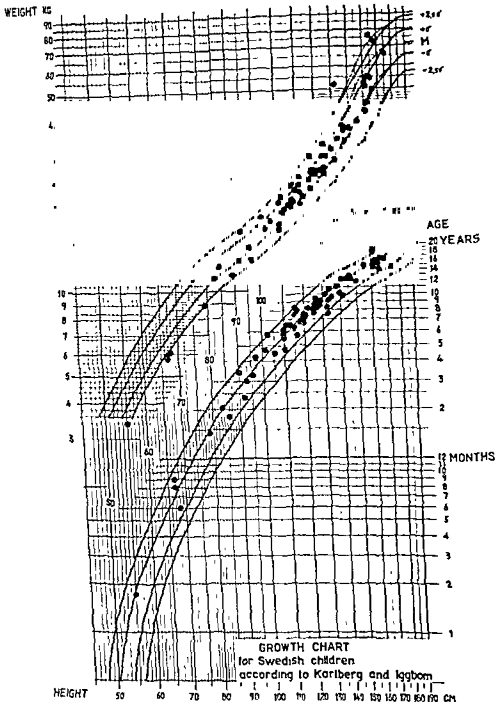


Fig 528 —Physical development in children with coarctation of aorta Height was normal in every case, and *only* two children were overweight (Relevant growth chart was kindly placed at our disposal by Drs P Karlberg and S Iggbom [374, 375])

wide patent ductus arteriosus with a large left to right shunt and pulmonary hypertension.

In an additional two cases, in which there was no complicating anomaly, cardiac symptoms appeared during the first year of life; a brief account follows.

Boy (K.B. 501119).—Nothing abnormal was noticed during the first months of life. From 6 months onward, he suffered from dyspnea. At this time, a roentgenologic examination because of pneumonia disclosed enlargement of the heart. At 11 months of age, he was admitted for the first time to the pediatric clinic of our hospital, because of a fresh respiratory tract infection. Coarctation of the aorta was diagnosed on this occasion. The blood pressure was 120/60 mm Hg in the arms and was not measurable in the legs. Only a faint systolic murmur was audible over the aortic area Loose râles were heard over both lungs. The roentgenologic appearance was characterized by considerable enlargement of the left atrium and left ventricle (see Fig. 545). During the following year, the heart increased greatly in size and right ventricular hypertrophy was superadded A precordial bulge had developed. It was therefore decided to operate at this early age. Cardiac catheterization and angiocardiography were performed preoperatively No shunts were present, but there were raised PCV pressure (13 mm Hg) and considerable pulmonary hypertension (60/32 mm Hg), which were attributed to left ventricular failure. The ECG showed marked left ventricular hypertrophy, but also signs of right ventricular hypertrophy and an incomplete right bundle-branch block. The P waves in leads I and II were notched and broad After operation, rapid improvement took place and the heart decreased in size

Boy, (A A 540310) —He progressed normally during the first months of life, then gained poorly in weight An acute exacerbation occurred in conjunction with a respiratory tract infection at 6 months of age He suffered thereafter from dyspnea Roentgenologic examination showed great enlargement of the left ventricle and left atrium (see Fig 543) The blood pressure in the arms was 130 mm Hg and in the legs 80 mm Hg (flush method (284)) A systolic murmur (grade 3) was audible over the pulmonary area and apex The ECG showed considerable left ventricular hypertrophy, and notched and broad P waves in leads I and II. Cardiac catheterization was performed As soon as the catheter entered the right atrium, paroxysmal tachycardia started, it was stopped by the administration of 2 ml of Cedilanid Consequently, the pressure in the pulmonary circulation was not recorded before digitalization. It nevertheless lay at the upper limit for the normal range of variations (31/8 mm Hg). The PCV pressure was normal (5 mm Hg).

The fact that cardiac symptoms may appear at an early age has been stressed by Gross (306), among others. He states that they are most prominent during infancy and then regress successively, so that later development of the patient is normal. Lang and Nadas (407) have shown that patients with heart failure during the first months of life respond very well to medical therapy, provided that digitalization is adequate and diuretics are given. In our experience, the symptoms may show some progression even after the first year of life. The reason for this early appearance of cardiac symptoms has not been established. The degree of stenosis cannot be the decisive factor, since many of our asymptomatic patients had atresia or extremely severe stenosis

According to Bahn *et al.* (33), left ventricular failure develops when the ductus arteriosus closes in those cases *in which* it is distal to the coarctation and collaterals are lacking (see p. 582). The development of the collateral circulation presumably plays a considerable role in decreasing the burden on the left ventricle. The collaterals appear to increase during early childhood. The left atrial enlargement which we have observed in young children and which has later regressed (see p. 600) is also an indication of a decrease in the load on the left ventricle during this period. Obviously, complicating cardiac diseases increase the risk of early heart failure. Endocardial fibroelastosis may occur in combination with coarctation of the aorta (77).

Physical Working Capacity

The physical working capacity is generally good in children with uncomplicated coarctation of the aorta The well developed collateral circulation ensures a good blood supply to the lower part of the body, and the coarctation does not limit physical activity. Even adult patients may have good

working capacity Thus a 20-year-old man (not included in our series) was able, in an exercise tolerance test on a bicycle ergometer, to perform work amounting to 1,200 kgm per minute, in a steady state with a pulse rate of 180 beats per minute, with no fatigue of the leg muscles.

Physical Signs

The most important sign in coarctation of the aorta is the difference between the blood pressure in the upper and in the lower part of the body. Blood pressure measurements in the arms and legs are not always performed routinely at health check-ups at schools and child welfare centers. Palpation of the femoral artery should therefore always be performed. This is a simple and

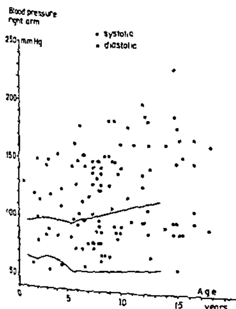


Fig 529.—Coarctation of the aorta Systolic (filled circles) and diastolic (open circles) blood pressure, measured with a cuff, in the right arm [illegible]
nots [illegible]
pre [illegible]
Nac [illegible]

rapid method of examination which has as great a justification as auscultation of the heart, it will reveal all cases of coarctation of the aorta with the exception of those with a fetal type of circulation. The pulsations in the femoral and dorsales pedes arteries are extremely faint or impalpable. On the other hand, the blood pressure in the upper

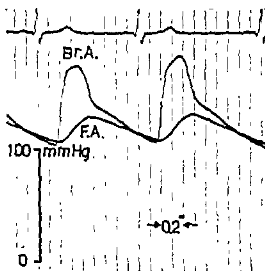
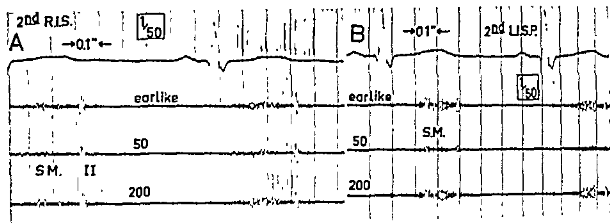


Fig 530.—Coarctation of the aorta Man, aged 20 (B B. 360902). Intra-arterial pressures, measured through catheters in brachial and femoral arteries. Note the small pulse amplitude in the femoral artery.

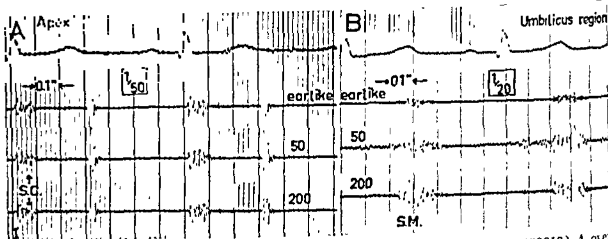
part of the body is not necessarily markedly raised in children (Fig. 529). After 10 years of age, there is a tendency to more marked hypertension Both systolic and diastolic pressures are raised, but the pulse amplitude in particular is greater than normal. The mean pressure is only inappreciably lower in the legs than in the arms, but the pulse amplitude is extremely low (Fig. 530). Werko *et al.* (695) found that the renal blood flow was only slightly decreased, an observation indicating that perfusion is satisfactory distal to the coarctation.

In one of our cases, the pressure was higher in the right arm than in the left; atresia was present at the site of origin of the left subclavian artery. In another case (J B. 490905) no pulsations were palpable in the legs and only faint pulsations in both arms, whereas the carotid artery on both sides exhibited strong pulsations. No blood pressure was measurable in the extremities,



... 531 —Phonocardiograms in coarctation of the aorta. Boy, aged 7 (B S 460607) A, over ...

... gree of amplification, other figures denote standard frequencies of the filters.



... 460612) A over ...

... mur (SM) is due to the coarctation and is transmitted caudally The murmur is late ... early diastolic but is not typical of a continuous murmur Boxed figures denote degree of amplification, other figures denote standard frequencies of the filters.

but on puncture of the carotid artery a systolic pressure of 140 mm Hg was recorded The coarctation was situated in the aortic arch, which had an abnormal course (see Angiocardiography, p 621). Both carotid arteries originated proximal to the coarctation, but the arteries to the arms were given off distal to it A similar case has been described by McGregor and Medalie (453).

In rare cases, the pressure may be higher in the left arm, i e., when the right subclavian artery originates distal to the coarctation and the left subclavian artery proximal to it (98). Palpation of the abdominal aorta should never be omitted, in view of the possibility of a low-lying coarctation Our series includes one case of coarctation of the abdominal aorta. No pulsations were palpable in the epigastrium The constriction started slightly below the diaphragm, and extended to the origin of the renal arteries

Apart from the difference between th

lood pressure in the upper and lower parts f the body, the physical signs are usually parse in children. Subcutaneous collaterals n the back or abdomen were observed in only four of our cases.

On the other hand, all of our patients had a faint to moderately loud systolic murmur, usually localized to the first or second intercostal space, sometimes to the left of the sternum and sometimes to the right as well The murmur was often transmitted to the back, and was then most distinct over the interscapular region (Fig. 531). The intensity is often greatest at the end of systole, and the murmur sometimes ends after the second sound, even when the ductus arteriosus is closed A typical continuous murmur was audible in only 10 of our 22 cases with a patent ductus.

The murmur may be extremely faint, particularly in infants We have had the opportunity of following one case from the neonatal period until 4 years of age No murmur was audible until after the patient was 1 year old

The blood flow through the coarctation cannot be the cause of the murmur in every case. Atresia of the aortic isthmus is often present, and the murmur is audible even in these cases. Complicating anomalies and the blood flow through the collaterals may be responsible for a murmur (307) This explains the great variation in the localization and nature of the murmur In coarctation of the abdominal aorta, the murmur is audible over the abdomen (355) This observation may, however, also apply in coarctation at the usual site, slightly distal to the origin of the left subclavian artery (Fig 532) The murmur is probably caused by the blood flow through the coarctation, and is transmitted to the abdominal aorta In our case of constriction of the abdominal aorta a continuous murmur was heard cephalad to the umbilicus (Fig 533)

In the cases complicated by aortic or valvular disease or ventricular septal defect, physical findings typical of these malformations were present

An apical diastolic murmur was heard in five cases without associated malformations. This murmur has been interpreted by many authors as a relative mitral stenosis due to the dilatation of the left ventricle and atrium. In the opinion of Wood (713), the murmur may be due to slight thickening of the mitral cusps.

When hypertension is fairly severe, the second sound is accentuated (Fig 532). Dilatation of the ascending aorta was the probable cause of the early systolic sound (*claquement aortique protosystolique*

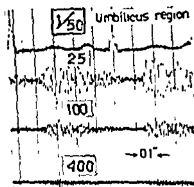


Fig 533.—Phonocardiogram in coarctation of the abdominal aorta. Girl, aged 12 (L.W. 450919). A continuous murmur was recorded in the umbilical region. Boxed figures denote degree of amplification; other figures denote standard frequencies of the filters.

(432)) heard over the aortic area or the apex in 12 of our cases (Fig. 532, A). There is a great similarity between the *claquement aortique protosystolique* in these cases and the early systolic sound in cases of dilatation and hypertension of the pulmonary artery (see p. 419). In 13 of our cases, the cardiac impulse was resistant and increased in breadth, as an expression of left ventricular hypertrophy.

Retinal changes of the type described by Granstrom (297) in adults are seldom present in children Only four of the patients with severe hypertension had slight retinal changes in the form of narrow, tortuous arteries

ELECTROCARDIOGRAPHY

The electrocardiogram was normal in 23 of the 53 cases with isolated coarctation or

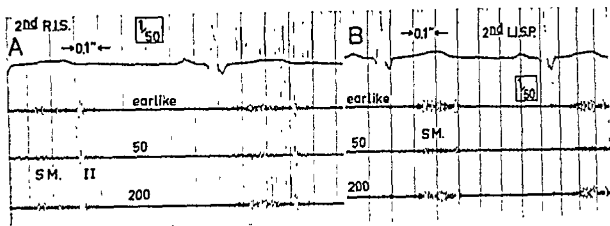
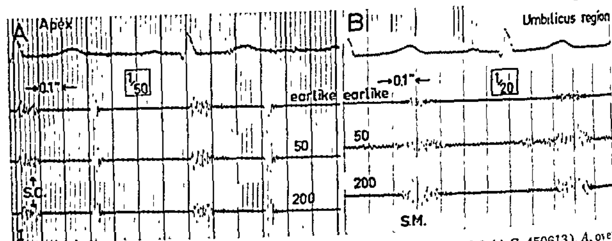


Fig. 531.—Phonocardiograms in coarctation of the aorta. Boy, aged 7 (B S. 460607). *A*, over 2nd right interspace (*RIS*) Fairly faint systolic murmur (*SM*) and split second sound *B*, over ... Murmur on the patient's back is of at least the same in... d by the collateral circulation. Boxed figures denote de... e standard frequencies of the filters

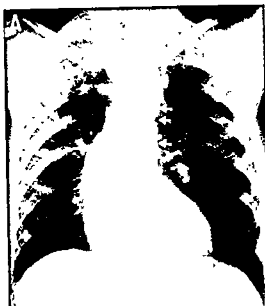


... coarctation of the aorta Boy, aged 9 (A.C. 450613). *A*, over ... (*SC*) is re... lic mur... olic and amplification, other figures denote standard frequencies of the ...

but on puncture of the carotid artery a systolic pressure of 140 mm Hg was recorded. The coarctation was situated in the aortic arch, which had an abnormal course (see Angiocardiography, p. 621) Both carotid arteries originated proximal to the coarctation, but the arteries to the arms were given off distal to it A similar case has been described by McGregor and Medalie (453)

In rare cases, the pressure may be higher in the left arm, i.e., when the right subclavian artery originates distal to the coarctation and the left subclavian artery proximal to it (98). Palpation of the abdominal aorta should never be omitted, in view of the possibility of a low-lying coarctation Our series includes one case of coarctation of the abdominal aorta. No pulsations were palpable in the epigastrium The constriction started slightly below the diaphragm, and extended to the origin of the renal arteries

Apart from the difference between the







535 —Coarctation of the aorta Boy aged 9 (A.C. 450612) ...

... mammary arteries (arrow in B)

with associated patent ductus arteriosus without any appreciable shunt. Signs of left ventricular hypertrophy were present in 13 cases. In these cases a pathologic P wave, indicating enlargement of the left atrium, was often seen. When the load on the left ventricle decreases after operation, normalization of the ECG takes place and may occur very rapidly (Fig. 534). In two patients there was combined left and right ventricular hypertrophy, and these patients had marked pulmonary hypertension as a manifestation of left ventricular failure In one patient complicating mitral incompetence was, however, present

The high incidence of right bundle-branch block is noteworthy (726) It was present in 17 of our cases In two cases there was a complete right bundle-branch block and in another two a prolongation of the P-R interval

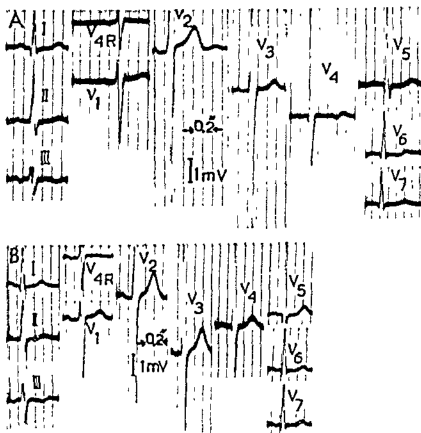


Fig. 534.—Electrocardiogram in coarctation of the aorta. Boy, aged 16 (O.E. 390318). *A*, before operation: inversion of T waves in V_4 and slight depression of T waves in V_{6-7}. *B*, 18 days after operation: the ECG is normal

ROENTGENOLOGIC EXAMINATION

In the majority of cases of coarctation of the aorta, the roentgenologic appearance is of such a nature that a diagnosis can be made without resort to special examination.

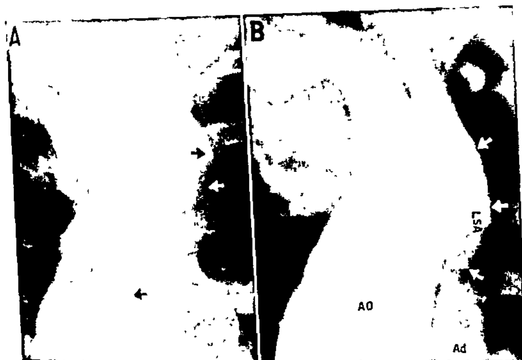
We made a roentgenologic examination of 95 patients: 61 children and 34 adults

The appearance found in adults, with deformation of the superior part of the mediastinum, dilatation of the ascending aorta, dilatation of the poststenotic segment of the descending aorta, enlargement of the left ventricle, notching of the ribs, and dilatation of the internal mammary arteries, is to a great extent characteristic in children as well (Figs 535 and 536). The changes in the appearance caused by the collaterals are, however, less prominent and cannot, as a rule, be demonstrated at all during the first years of life.

depending on the extent to which this segment of the vessel is retracted toward the mediastinum If the subclavian artery is wide and there is poststenotic dilatation of the descending aorta and the constricted segment is curved forward, as is common, a double-curved, partly overlapping outline of characteristic appearance is seen in the mediastinum (Figs. 535, 538, and 539)

...tation was slight or lacking. ...ie ...n- ...il- ...is not pathognomonic of coarctation of the aorta.

It is generally possible on an ordinary roentgenologic examination to determine



the right accentuates the impression in the esophagus at the level of the left main bronchus (bottom arrow in A). *Ad*, descending aorta, *AO*, ascending aorta, *LSA*, left subclavian artery.

Only in rare cases does the distal segment of the aortic arch have a common outline with the mediastinal pleura (Fig 546) Figley (252) has pointed out that there may be a break in the mediastinal outline at the level of the transition to the descending aorta This is caused by retraction of the segment of the vessel at the stenosis (Fig 540) In our series, it was observed in both adults and children, particularly when the

the site of the stenosis by identifying the upper borderline of the descending aorta. Since in this segment of the ...

most suitable, so that the vessel is depicted over the spinal column (Fig 538), this has already been stressed by Wolke (710). Part of the subclavian artery can then be visualized as well, which is seldom possible in

The most constant and, at the same time, the most specific feature is the deformation *of the superior part of the mediastinum.* Among the publications on this subject, mention may be made of those of Schatzki and Hallermann (578), Ernstene and Robins (241), Wolke (710), Gladnikoff (280), Fleischner (258) and Bruwer and Pugh (117). On the basis of 75 of his own cases, Figley (252) *has given a comprehensive* description of the roentgenologic features, seen at the normal site of the aortic arch or slightly below it. It is caused by the dilated left subclavian artery (Figs. 535-537), but this vessel and the aortic arch may occasionally be superimposed in the mediastinal outline. The aortic arch may sometimes have a separate outline, medial to the lateral borderline of the subclavian artery (Figs 535 and 537).

The appearance of the superior segment of the mediastinal outline on the left side is



Fig. 536.—Coarctation of the aorta Boy, aged 6 (K.P. 460131). Mediastinal changes are less marked than in Figure 535 Slight notching of the 4th–6th right ribs and 7th–8th left ribs

with special reference to the anatomy of the mediastinum.

The details of the mediastinal deforma-tion are subject to considerable variations and may be difficult to analyze. This is because the vessels of the mediastinum are visualized only to the extent to which they form an outline against the lung.

In 68 cases, we made a parallel study of the roentgenograms and the angiograms, taken in frontal and lateral projections. The absence of any distinct aortic arch segment is due partly to the narrowness of the arch and partly *to its abnormal course. A* definite prominence is nevertheless usually determined by the course and width of the subclavian artery When this vessel is dilated, *as is often* the case, the mediastinum is broader than normally. The mediastinum may have a convex, straight, concave, or S-shaped outline, according to the width and course *of the vessel in* question. However, hypoplasia of the left subclavian artery, demonstrated on angiocardiography and operation in one of our cases, could not be predicted on the basis of the mediastinal picture

The transition into the descending aorta *is often marked* by an indentation in the mediastinal outline. Its appearance varies,

the lateral projection. It is not, however, possible to determine the exact length and shape of the stenosis, owing to the displacement of the distal segment of the aortic arch This is because it is depicted more or less axially in the frontal or oblique projection, even if the segment happens to form an outline against the lung. As a rule the descending aorta can be identified directly lateral to the spinal column, as a faintly bulging outline due to poststenotic dilatation contributes to this shift (Figs. 538 and 539) It should, however, be borne in mind that the same local deviation of the esophagus may occur normally, although it is then less marked No definite displacement of the esophagus at the level of the dilated left subclavian artery was demonstrated in our series. The mediastinal deformity was not visible in our infants, nor did it appear in a 13-year-old boy with a greatly dilated pulmonary artery associated

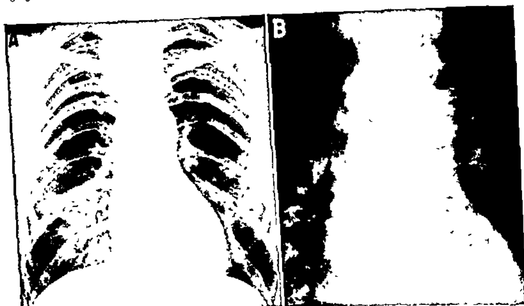


Fig 539.—Coarctation of the aorta Man, aged 29 (T B 240602) Considerable poststenotic dilatation of descending aorta (arrow in *B*), slight dilatation of ascending aorta and left subclavian artery, inappreciable enlargement of left ventricle, plentiful notching of ribs

tion of the vessel and a slightly curved course. In one of our cases no such outline of the descending aorta was visible. Angiocardiography showed that the entire vessel was situated anterior to the spinal column, closer to the midline than in other cases

The interpretation of the mediastinal changes may be facilitated by a study of the course of the esophagus The indentation made in it by the aortic arch is often lacking or slight If there is considerable displacement of the stenosed segment, the esophagus sometimes deviates forward and to the right at the level of the carina and the left main bronchus The poststenotic dila- patent ductus arteriosus and heart failure.

The ascending aorta may be of normal width or dilated. The dilatation is most distinct in the suprabulbar segment It may, as illustrated in Figure 535, be considerable and of the same appearance as in valvular or subvalvular aortic stenosis

Enlargement of the left ventricle is common and occurred in slightly more than half of our cases. In our series, the enlargement was generally moderate Gross enlargement was present in the infants, in two children with associated patent ductus and a large left to right shunt, in one of them with heart failure as well, and in two ado-



Fig. 538.—Coarctation of the aorta Man, aged 23 (B.S. 30). Considerable poststenotic dilatation of the descending aorta (arrow in C), most distinct in the oblique projection (D) Left subclavian artery (right arrow in A) overlaps the aortic arch (left arrow in A). Typical displacement of the esophagus (arrow in B), dilatation of the ascending aorta, no notching of the ribs

We made roentgenologic examinations of r infants with coarctation of the aorta. e of them showed considerable enlargement of the heart during the first day of e A reduction in size took place during e following days, succeeded by an increase This course is a normal occurrence 998a) The left atrium was not enlarged. he initially increased vascularity of the ings gradually regressed No mediastinal eformity could be demonstrated. On examnation one and three years later, the heart was still enlarged and the left ventricle formity could be visualized and the ascending aorta was not prominent. The vascularity of the lungs was increased, presumably as a manifestation of pulmonary stasis At 14 months of age, as well as one year later, the enlargement of the left atrium was still as distinct A shallow indentation in the mediastinal outline marked the site of the coarctation In the fourth patient (Fig 543), who was investigated at 4 months of age, the left atrium was greatly dilated and the left ventricle exhibited considerable enlargement and in-

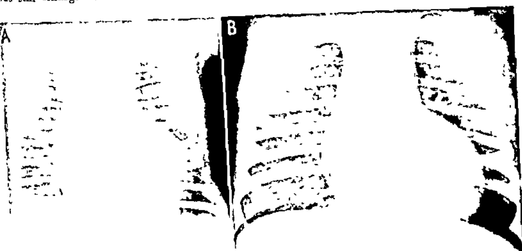


Fig 541.—Coarctation of the aorta Girl (C N 521030) Shape of heart at *A*, 1 day, *B*, 1 year On latter examination, gross hypertrophy of left ventricle has appeared, which gives the heart the appearance typical of the disease at this age

exhibited the typical hypertrophy (Fig 541) The left atrium was dilated At the last examination, distinct deformity of the aorta was seen No erosion of the ribs could be demonstrated

Another patient was examined at 3 weeks of age A complicating ventricular septal defect and left to right shunt was present The heart was considerably enlarged, all its chambers being involved, not least the left and right atrium No mediastinal deformity was displayed and notching of the ribs was not present Of the remaining two, one was examined for the first time at barely 3 months of age The heart was moderately enlarged and the left atrium was distinctly dilated (Fig 542) No mediastinal decreased curvature The same appearance was found in a 1-year-old child (Fig. 545), who was operated on at 18 months of age, owing to heart failure This typical roentgenologic appearance is important from the prognostic viewpoint, since it implies severe overloading of the left ventricle, which makes it necessary to consider early operation

We had eight cases in the 1 to 5 year age group The heart was large in all of them, in six, enlargement could be attributed to dilatation of the left atrium and a large left ventricle (Fig 544). In an additional case, all chambers of the heart were dilated, the left atrium to the greatest degree (Fig.

lescents with complicating aortic and mitral incompetence, respectively. The change in the shape of the left ventricle was not uniform, and the appearance and position of the apex varied. In some of the cases its outline was definitely curved.

Ever since Roesler (561) and Railsback and Dock (546) pointed out that notching



Fig. 540.—Coarctation of the aorta Woman, aged 23 (V S 30) Discontinuity of mediastinal outline corresponding to segment at level of the coarctation (upper right arrow). Large impression in the esophagus (left arrow) Contour of the descending aorta is indicated by the lower right arrow

of the ribs appears on the roentgenogram in coarctation of the aorta, such notching has been one of the classic findings in this malformation Later experience (252) has nevertheless shown that notching of the ribs may occur in other conditions and that it is therefore not specific for coarctation of the aorta. Moreover, this feature can never be demonstrated in small children. It may also be lacking in adults, a fact which does not necessarily imply that the stenosis is less severe from the functional viewpoint. The notches usually occur bilaterally on the third to eighth ribs; they may be multiple or single and are on the lower margins of the ribs, corresponding to the upper margin of the costal sulcus. In our series, notching of the ribs was found in retrospect as small irregularities in the outline of the ribs in a few patients 2 to 3 years of age, but such irregularities could not be visualized distinctly in these cases before the age of 4 or 5. Unilateral notching of the ribs was not present in any of the cases.

Another characteristic roentgenologic feature has been described by Odman (514). He has pointed out that the tortuous and dilated internal mammary arteries are projected in the lateral view as soft tissue densities behind the sternum. These arteries are not visible as distinctly in children as in adults. In one case we were able to identify them in a 4-year-old child, but they could be seen more frequently only after the age of 10 (see Fig. 549).

Indentations in the esophagus caused by dilated mediastinal vessels belonging to the collateral circulation (226, 306, 587) were seen in only one of our cases.

Some of the variations which occur in the roentgenologic appearance of the heart and the aorta are dependent on the varying age of the patients. This applies in particular to the development of the collateral circulation and to the size of the left atrium and left ventricle.

Few accounts have been given of the roentgenologic appearance in coarctation of the aorta during the first weeks of life. Buchs (118) followed the clinical course and the change in roentgenologic appearance in six cases from the neonatal period until 3 to 5 months of age. He demonstrated, for example, that there is an initial stage of decompensation, with generalized enlargement of the heart and pulmonary stasis. This is followed by regression of the cardiac dilatation, together with a successive increase in the prominence of the ascending aorta and a decrease in the prominence of the pulmonary artery.

545) In several cases the heart volume was increased to more than twice the normal. The enlargement of the left atrium could not be ascribed in any case to coincident mitral valve disease In one case with associated patent ductus arteriosus and a large left to right shunt, there was reason to assume that the abnormal flow contributed to enlargement of the atrium. In one case the left atrium had decreased distinctly in size within three months after operation for coarctation Four years later, no enlargement of the atrium was present and the heart volume was normal. In two other cases no definite regression of left atrial dilatation could be observed one and one-half and three years, respectively, after operation

Small erosions of the ribs were observed in two 3-year-old children. A typical deformity of the mediastinal outline was not seen before 2 years of age This sign has only occasionally been observed at an earlier age (476)

The age group 6 to 15 years includes 42 patients No enlargement of the heart was present in 24 of them. Moderate enlargement was found in most of the others. Considerable enlargement was present in four patients with associated aortic or mitral incompetence (Fig 547) and with associated patent ductus with a large left to right shunt, with or without heart failure (Fig. 548). In all these complicated cases, the left atrium was considerably enlarged. In 18 cases inappreciable or moderate enlargement was present, and in the remaining 20 cases there was no dilatation of the atrium. In the case with associated mitral incompetence, the atrial enlargement was unchanged four years after operation. The internal mammary arteries appeared distinctly (Fig 549), and notching of the ribs was observed in slightly less than half of the cases The mediastinal deformity was, with few exceptions, typical in this group. In one case the coarctation was situated particularly high up (Figs 550a and 550b).

In 35 patients, all over 15 years old, the roentgenologic appearance was characteristic and in agreement with the descriptions of other writers. Definite enlargement of t

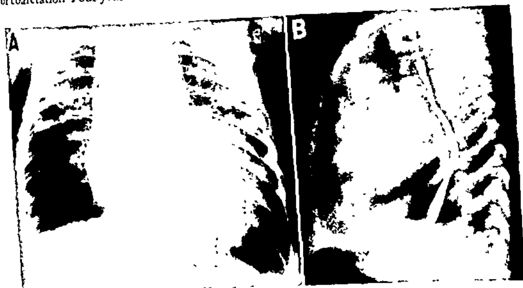


Fig 543.—Coarctation of the aorta Boy, aged 4 months (A.A 540310) Considerable increase in heart volume, dilatation of left atrium and ventricle in particular. The great hypertrophy and dilatation of the left ventricle is reflected in marked curvature of the left border, which we have otherwise seen only in atresia of the tricuspid valve Pronounced pulmonary stasis Aorta cannot be evaluated

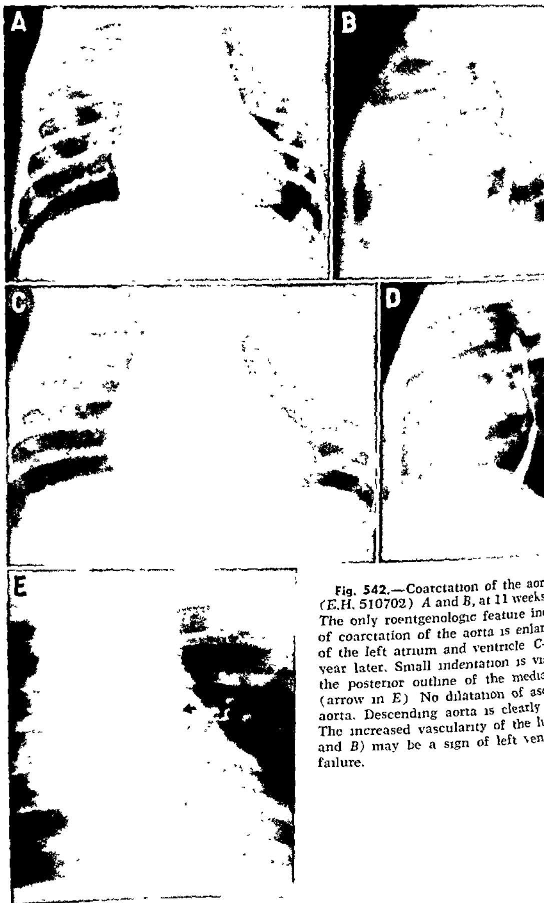


Fig. 542.—Coarctation of the aor
(E.H. 510702) *A* and *B*, at 11 weeks
The only roentgenologic feature in
of coarctation of the aorta is enlar
of the left atrium and ventricle C-
year later. Small indentation is vi
the posterior outline of the medi
(arrow in *E*) No dilatation of as
aorta. Descending aorta is clearly
The increased vascularity of the lu
and *B*) may be a sign of left ven
failure.





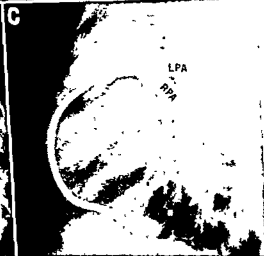


Fig 544b —Same case as in Figure 544a The enlarged left atrium (*B*) lifts the right branch of the pulmonary artery but does not markedly displace the secondary branches. In the lateral projection (C), the site of the latter is normal and the right branches run caudally to and in front of the left branches. *LA*, left atrium, *LPA* and *RPA*, secondary branches of left and right pulmonary arteries, *PA*, pulmonary artery

left atrium was present in one case, and only slight enlargement in 12.

Enlargement of the left atrium is thus a prominent feature in small children with coarctation of the aorta. The typical forward deviation of the esophagus at the level of the coarctation tends to accentuate the impression of atrial enlargement. We were therefore careful to bear this fact in mind in our interpretation Angiocardiography, performed through the left atrium or the pulmonary artery (Figs. 544 and 546b), nevertheless showed that the atrium was, in fact, enlarged. The enlargement, which was considerable when heart failure was present and unquestionable in the other cases, is presumably an expression of the impeded emptying of the left ventricle, owing to the fact that the collateral circulation is relatively undeveloped at this time This view is supported by other observations. When the collaterals were sparse but the left atrium was not dilated, a definite communication in the stenosis could be shown on angiocardiography and was confirmed at operation (Fig. 551) Dilatation of the left atrium is far less common and less marked in adults than in children. There is no reason to assume that the adults in our series represent a selection of patients who have never exhibited left atrial enlargement. It is, on the contrary, probable that dilatation of the left atrium in children gradually regresses, presumably as an expression of a decrease in the overloading of the left ventricle by means of improved collateral circulation



Fig. 544a.—Coarctation of the aorta Girl, aged 4 (A.Ö. 480727). The greatly enlarged left atrium overlaps the right and forms part of the right border of the heart (left arrows in *B*) Esophagus is displaced to the left (lower right arrow in ... ascending aorta is visible up to the middle arrow in *A* and upper right arrow ... gion of the coarctation Dilated subclavian artery (top ar... notching of ribs.

Electrokymography

Electrokymograms were recorded over the ascending aorta, the aortic arch (left subclavian artery), and the descending aorta in 10 cases The pulsations of the left atrium were studied in some older children and a few adults.

Pathologic tracings with a sinus-shaped appearance and a delayed upstroke were obtained on recording the aortic pulsations below the stenosis in four cases (Figs. 552–554) Taken over a segment still lower down, the curves were modified by an extra wave in early systole (Figs. 552 and

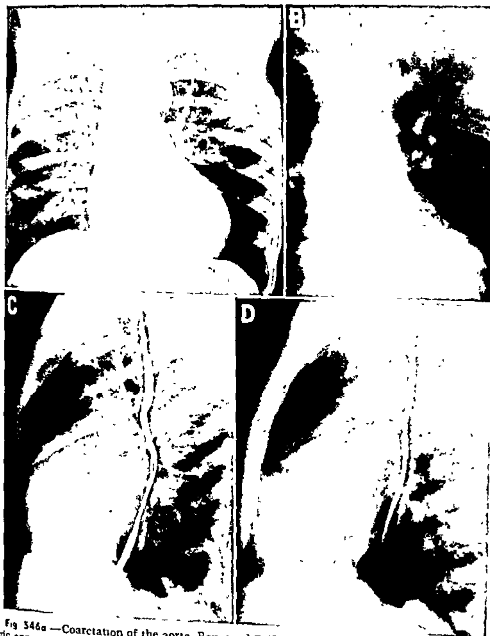


Fig 546a —Coarctation of the aorta Boy, aged 7 (B S 460607) . . .
logic appearance with d . . .
The marked bul
The subclavian
outline (see *I* in
B and D, posteroa-
aorta and

stasis and dilatation of ascending aorta *C* and *D*, at 19 months Considerable increase in heart volume, great enlargement of left atrium and forward displacement of the anterior wall of the thorax, marked pulmonary stasis *E* and *F*, one year later (eight months after . . . crease in heart volume, precordial bulge scarcely . . .
atrium, vascularity of . . .



Fig. 545.—Coarctation of the aorta. Boy (K B 501119), see Figure 558. *A* and *B*, at 12 months of age Increase in heart volume, with distinct enlargement of left atrium, pulmonary

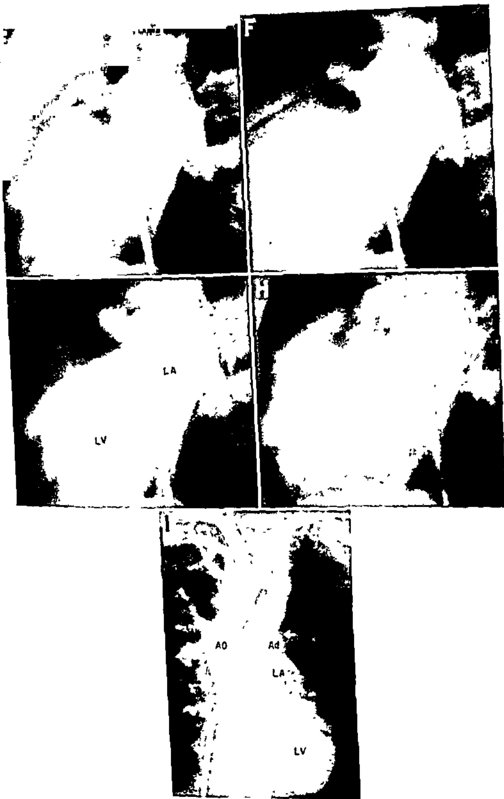


Fig. 546b (*cont.*)



Fig. 546b.—Same case as in Figure 546a Injection of contrast medium into the left atrium, which is large and does not empty to same extent as normally. (*H*, late ventricular diastole) Left ventricle has a thick wall and is distinctly dilated Moderately large quantity of residual blood (*B*) Small leakage of contrast medium to right atrium through the foramen ovale. Arrow in *I* points to the coarctation No poststenotic dilatation of descending aorta *Ad*, descending aorta, *AO*, ascending aorta and aortic arch, *LA*, left atrium, *LV*, left ventricle (*continued*)

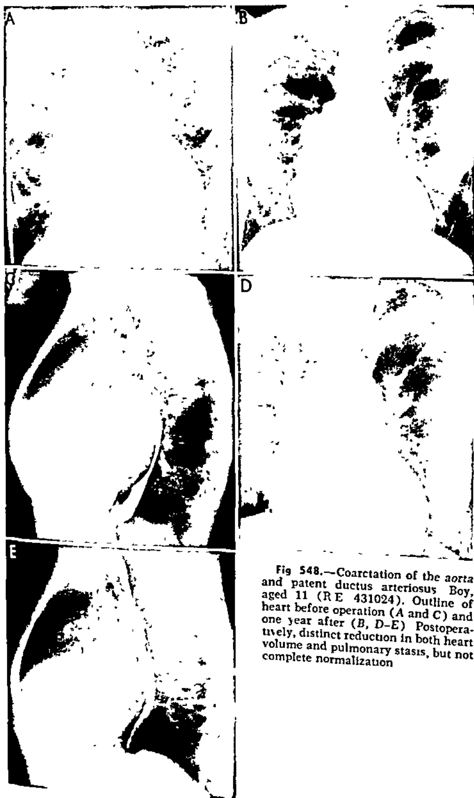


Fig 548.—Coarctation of the *aorta* and patent ductus arteriosus Boy, aged 11 (R E 431024). Outline of heart before operation (*A* and *C*) and one year after (*B*, *D–E*) Postoperatively, distinct reduction in both heart volume and pulmonary stasis, but not complete normalization



Fig. 547.—Coarctation of the aorta and aortic incompetence Boy, aged 15 (A.C. 420325) All pictures taken after end of injection Marked regurgitation from aorta (*AO*) to left ventricl (*LV*), which is large.



Fig 550a —Coarctation of the aorta Girl, aged 8 (E S 461008) Fairly high-lying coarctation (arrow in C) with considerable poststenotic dilatation Typical medial and forward displacement of constricted segment

553). In two other cases, such alterations were barely discernible. In the others, the configuration was the same in the electrokymograms recorded above and below the stenosis.

Several factors presumably constitute the hemodynamic basis of the alterations in the electrokymograms recorded over the poststenotic region. They are the altered shape



Fig. 549.—Coarctation of the aorta Boy, aged 9 (G.W 440127) The wide, tortuous mammary arteries are outlined on the inner aspect of the anterior thoracic wall (arrows).

of the pulse wave in the stenosis (396), the volume and type of the collateral flow and, possibly, the movements in the aortic wall produced by the impact of the pulse wave on the stenosed segment

Fluoroscopic observations showed that the pulsations in the descending aorta were usually small. This applied particularly in a comparison with the segment of the aorta and the left subclavian artery above the stenosis. These conditions can be demonstrated convincingly with roentgenkymography (412, 627). In cases with a fairly wide communication in the stenosis established on angiocardiography and at operation—but also in a few cases with total or subtotal stenosis—the pulsations were, however, demonstrated to have the same large amplitude on each side of the stenosis.

In the tracings of the ascending aorta and the aortic arch, the dicrotic wave was small in several cases. In the former curves, the incisura was low down on the descending limb (Fig. 553), presumably as an expression of the increased pulse amplitude in the prestenotic portion of the aorta. An electrokymogram with the same appearance was obtained in one case with associated incompetence of the aorta (Fig 555).

The left atrial tracings were normal In one case with coincident mitral valve disease, it was possible to rule out electrokymographically that it was due to stenosis. On the other hand, it was not possible to establish the existence of mitral incompetence, which was strongly indicated by the clinical findings (Fig 556).

In coarctation of the aorta with a fetal type of circulation, the pulmonary artery tracing had the appearance characteristic of pulmonary hypertension (Fig. 557). The aortic electrokymograms were, on the contrary, normal.

CARDIAC CATHETERIZATION

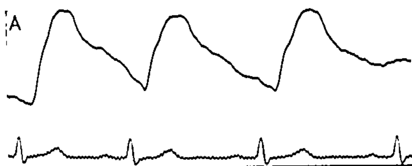
The diagnosis of coarctation of the aorta is made on the basis of the physical findings. Consequently, cardiac catheterization may appear to be superfluous. We nevertheless made this examination in all but 10 of our cases, on the following indications (1) To ascertain whether there was any complicating intracardiac malformation in clinically atypical cases (2) To determine the pulmonary artery and pulmonary capillary venous pressures in cases with signs or symptoms of left ventricular failure. (3) In order to perform angiocardiography with injection into the pulmonary artery, this was the main indication for catheterization of the right side of the heart.



(R.V. 451228). Moderate hypertrophy of left
ilatation of the aorta, no demon-
cation in the stenosis.



Fig. 550b.—Same case as in Figure 550a. The coarctation (arrows) lies directly below the origin of the left subclavian artery. Moderately well-developed collateral circulation. Contrast medium injected into pulmonary artery *Ad*, descending aorta; *AO*, ascending aorta



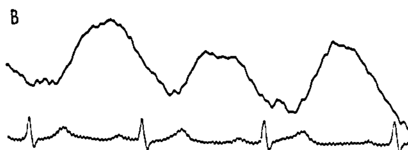


Fig. 554.—Electrok

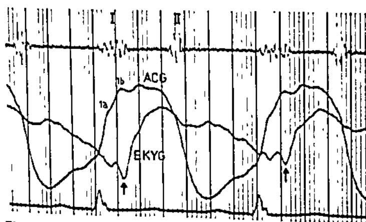


Fig. 555.—Electro
rtic incompete
ectrokymogram
und, ACG, ape
milunar valve.

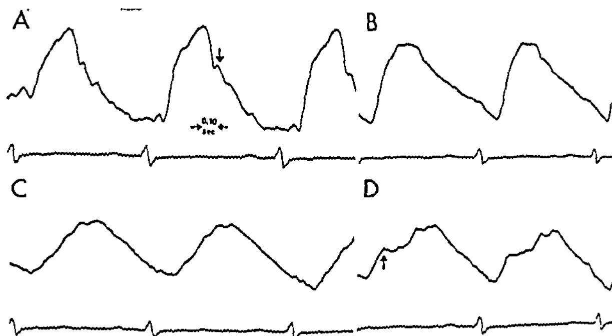


Fig. 552.—Electrokymograms in coarctation of the aorta. Man, aged 24 (S J. 270125). Time marking, 0.10 sec between thick lines. *A*, ascending aorta. Onset of upstroke at normal time. Dicrotic wave small (arrow). *B*, aortic arch. Suggested reduction of dicrotic wave. Onset of upstroke, 0.14 sec after Q wave. *C*, descending aorta directly below the stenosis. Curve is sinus-shaped. Incisura and dicrotic wave are obliterated. Onset of upstroke, 0 18 sec after Q wave *D*, still farther down As in *C*, but modified by a positive wave (arrow) with onset 0.15 sec after Q wave.

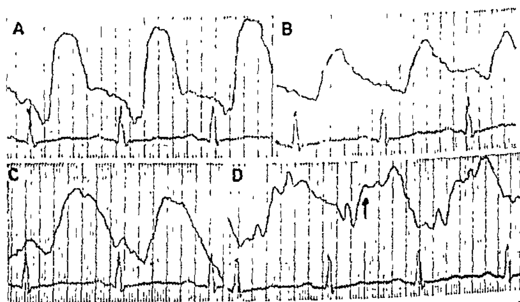
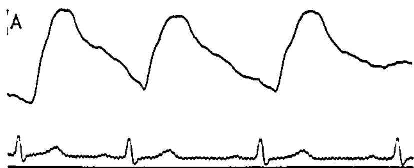


Fig. 553.—Electrokymograms in coarctation of the aorta. Girl, aged 17 (B.T. 370611). *A*, ascending aorta Incisura far down and dicrotic wave small Onset of upstroke at normal time. *B*, aortic arch (left subclavian artery) Incisura and dicrotic wave lacking in prominence. *C*, descending aorta directly below the stenosis Onset of upstroke is late—0 21 sec after Q wave. Summit of curve is displaced toward diastole Tendency to a sinus shape *D*, still farther down. Positive wave (arrow) with a high gradient in first part of upstroke. Irregular small deflections are superimposed on the curve before the crest is reached in late diastole



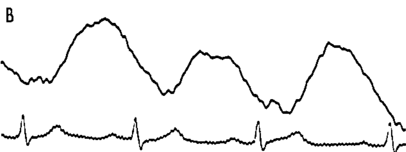


Fig 554 —Electr... diastole The c...

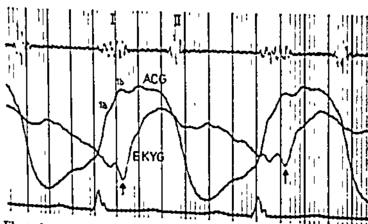


Fig 555.—Electrokymogram of ascending aorta in ... of the aorta and associated aortic incompetence Boy, agec ... electrokymogram at normal ti ... Onset of upstroke in aortic ... wave I, 1st sound, II, 2nd sound, ACG, apex cardiogram ...ricular valve, 1b, opening of semilunar valve

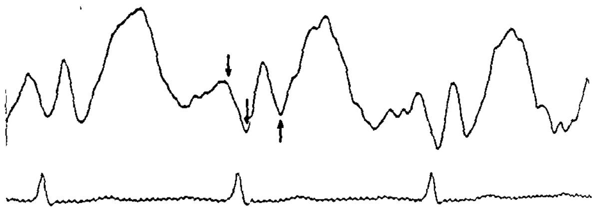


Fig. 556.—Left atrial electrokymogram in coarctation of the aorta with associated mitral incompetence. Girl, aged 9 (*M.L. 431114*). Atrial systole (between left-hand arrows) is not prolonged—0.08 sec Upstroke, *corresponding* to the filling phase in systole (right arrow), is at the normal time—0 18 sec after Q wave. Tracing has the normal appearance.

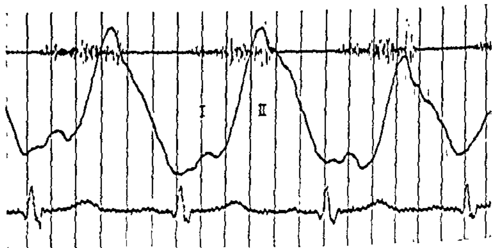


Fig. 557.—Electrokymogram *of pulmonary* artery in coarctation of the aorta with fetal type of circulation Boy, aged 12 (L A. 420531). PCG over pulmonary area. Onset of upstroke late—0.17 *sec after* Q wave; slope continuous to the summit, at the end of systole Incisura and dicrotic wave poorly *defined and lie high* up on the descending limb They are synchronous with the 2nd pulmonary sound. *I*, 1st sound; *II*, 2nd sound

In coarctation of the aorta, complicating malformations are usually localized to the left side of the heart, and particularly to the aortic orifice No information is therefore given by catheterization of the right side of the heart

Mild pulmonary hypertension with a mean pulmonary artery pressure between 20 and 30 mm Hg occurred fairly often Any more marked rise in pressure (mean pressure between 40 and 90 mm Hg) was observed only in patients with associated malformations, such as a wide patent ductus and mitral or aortic valve lesions In seven cases in which coarctation was present as an isolated malformation, the PCV pressure was slightly raised (12 to 16 mm Hg at rest) All these patients had distinct enlargement of the left atrium.

A patent ductus arteriosus is often present in combination with coarctation of the aorta However, as a rule it is so narrow that it lacks any functional importance A left to right shunt could be demonstrated in only 17 of our 22 cases in which the ductus was found to be patent at operation Only in three cases was the difference between the oxygen content of the right ventricle and the pulmonary artery more than 1 volume per cent.

ANGIOCARDIOGRAPHY

In view of the indications for operation, angiocardiographic examination in coarctation of the aorta has been regarded in some quarters as unnecessary In Sweden, it is performed routinely, since the surgeons wish to obtain exact information about the anatomy of the stenosis before operation It is of paramount importance to diagnose the low-lying, possibly multiple stenoses, as well as the elongated type of coarctation or atresia, in which a graft is required at operation

A detailed account of the roentgenologic anatomy in coarctation of the aorta has been given by Jonsson, Brodén, and Karnell (370) Their series consisted of 36 patients, all over 6 years old and all examined by thoracic aortography On the basis of this material, they illustrated the different anatomic variants and also suggested the following classification for an appraisal of the cases from the surgical viewpoint:

Group I: Distal part of the arch fairly long and wide
Group II: Distal part of the arch fairly long but narrow
Group III: Distal part of the arch short
Group IV: Distal part of the arch entirely without a lumen—atresia

This classification is also fully applicable in younger children

The results of angiocardiography depend greatly on sufficient density of the contrast medium in the aorta.

When *intravenous injection* is used, the density is not infrequently unsatisfactory, owing to dilution of the contrast medium during passage of the aorta (574, 585).

Acceptable results which are not, however, invariably satisfactory can be anticipated with *injection into the main trunk of the pulmonary artery or the right ventricle* (Fig 551). Using this method, we were able to obtain detailed information about the appearance of the stenosis and the collateral circulation in 26 of 43 children. Satisfactory depiction of the anatomy of the aorta but somewhat incomplete information regarding the extent of the collaterals were obtained in 10 cases. In the remaining seven cases, the angiocardiograms were of poorer quality, so that the collateral vessels and the deformity at the site of coarctation were to some extent incompletely outlined.

Some of the unsuccessful cases were among the first in our series to be examined, at a time when the technique was not yet fully developed to give adequate information The others consisted partly of cases with considerable dilatation of the left atrium, in combination with left ventricular strain or patent ductus arteriosus with a left to right shunt. In three patients, all adolescents, dilution of the contrast medium on passage through the pulmonary circuit was presumably responsible for unsatisfactory opacification of the aorta, despite absence of any noteworthy atrial en-

largement or stasis. It is probable that such conditions are more generally applicable in examination of adults.

In *thoracic aortography*, the anatomy of the isthmus region can be seen in all its details, the collateral circulation can be visualized, and the course of filling of the descending aorta can be followed. Various techniques have been reviewed and illustrated by Jonsson (368). In 28 examinations we used the technique devised by Jönsson *et al* (370), consisting of introduction of a Cournand or Lehman catheter into the ascending aorta after exposure of the radial artery. It has been recommended that the tip of the catheter be placed in the middle segment of the ascending aorta. We have often preferred to allow the injection to start in its lower part, but at a safe distance from the coronary orifices. This is because the relatively fine caliber of the catheters (no 4–7) used in children makes it necessary to increase the pressure of injection. Consequently, the tip of the catheter may recoil appreciably during the injection, with a resulting risk of an overdose of the contrast medium to the cerebral vessels if the distance from the vessels of the neck is not sufficiently great.

As stated on page 124, we subsequently modified the technique and made the injections through catheters with lateral holes and a sealed tip As a rule, the contrast medium was then deposited in the lower part of the ascending aorta. We performed thoracic aortography mainly in adults and adolescents and less often in children. The youngest child to be investigated with this method was 18 months of age (Fig. 558).

In some cases a polyethylene catheter was introduced percutaneously into the radial artery according to Seldinger's method (588) and the tip of the catheter placed in the ascending aorta Catheters of this material are not at present as stiff as Cournand or Lehman catheters, it is therefore necessary to use a somewhat lower pressure of injection in view of the greater risk of troublesome recoiling

In one case the contrast medium was injected into the left atrium after the catheter introduced into the saphenous vein had been passed through a patent foramen ovale. The result was satisfactory. A very diluted flow of contrast medium into the pulmonary artery by means of a leak through the foramen ovale caused no essential difficulties in the interpretation

Retrograde brachial aortography was performed in two infants and one small child (Fig. 559). This method has been described by Castellanos and Pereiras (148, 149), and its value has been confirmed by many authors (3, 123, 379, among others) One of our infants examined with this technique represented coarctation of the aorta with a fetal direction of the circulation (see p. 640, Fig. 583).

Experience from 75 angiocardiographic examinations on 71 patients showed that the orifice could only exceptionally be visualized by means of passage of a contrast-filled jet through it (Figs. 560 and 561) In the large majority of cases, the maximal stenosis involved a short segment and had the appearance of a membrane. Specimens from the stenosed segment taken at operation and autopsy in three cases are shown in Figures 563–565. In most of the cases the aortic arch was curved downward, owing to the typical displacement of its distal segment. As a rule, the middle segment of the arch was narrower than normally and of the same width as the dilated left subclavian artery. The middle segment was wider in cases with a distinct communication in the stenosis (Figs. 551 and 561). The distance from the left subclavian artery to the systemic arteries on the right side was strikingly short. In one case the coarctation was situated in the aortic arch proximal to the left subclavian artery (Fig 562). The latter probably served as a collateral via the left carotid artery.

Filling of the descending aorta took place slowly. In approximately half of the cases, the contrast medium was observed in the descending aorta before opacification of the collaterals, indicating the presence of a communication in the stenosis In the remainder, the descending aorta



Fig 558 —Coarctation of the aorta ... aorto
satisf
tation
scending aorta scanty collateral circulation C and D, injection of contrast medium into pulmonary artery Great dilatation of left atrium, with very incomplete emptying C, early ventricular systole, immediately after the R wave in the ECG D, late ventricular systole (picture slightly enlarged) Ad, descending aorta AO, ascending aorta and aortic arch LA, left atrium, LSA, left subclavian artery

was not filled until after the collaterals had been visualized. Local poststenotic dilatation was generally less conspicuous in children under 7 years of age than in older children and adults. The degree of dilatation varied considerably (Figs 550, 551, 560 and 566–570). Caudal to this segment, the descending aorta was strikingly narrow in many cases.

With few exceptions, the collateral circulation was sparsely developed in children under 7 years of age Scapular anastomoses and the meshwork of vessels at the superior aperture of the thorax, corresponding mainly to cervical collaterals, could be demonstrated in most of the cases but were not as extensive as in older children and adults. The internal mammary arteries exhibited only moderate dilatation and slightly increased tortuosity. Jonsson *et al* (370) have described collateral arterial trunks connecting the intercostal arteries given off by the costocervical trunk and the superior part of the aorta (Fig. 571). In our series, they were far less extensive in children (Fig. 566) than in the majority of adults, in whom these vascular trunks and the mammary arteries formed a predominant part of the collateral circulation (Fig. 572).

The ascending aorta was dilated in more than half of our cases. The dilatation was most apparent in the segment above the aortic bulb. It was not especially marked in four patients with incompetence of the aortic cusps or in a patient with associated aortic valvular stenosis (see Fig. 609, p 668, Aortic Stenosis) In one case with complicating mitral incompetence, the ascending aorta was not dilated, but was possibly somewhat narrow (Fig. 573).

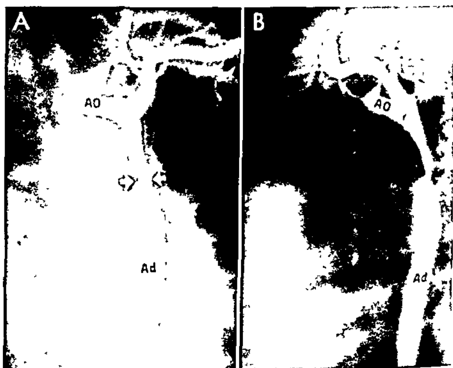


Fig. 559.—Coarctation of the aorta. Boy, aged 1 month (B.E 560801). Contrast medium injected into brachial artery through catheter inserted in this vessel. The coarctation (arrows), about 2 cm caudal to origin of left subclavian artery, has a lumen about 1 mm wide. Poststenotic dilatation of descending aorta (*Ad*) *AO*, aortic arch.

Three sinuses of Valsalva were visible in all but one of the patients, in whom only two sinuses could be identified with certainty (Fig 574)

A patent ductus arteriosus of small caliber was found in 22 cases at operation for stenosis of the isthmus. It had been demon-



Fig 560 —Coarctation of the aorta (type 1) Man, aged 29 (T B 240602) Membranous stricture with [illegible]
typical d [illegible]
larly do [illegible]
mamma [illegible]
aorta, J, jet, MI, internal mammary arteries

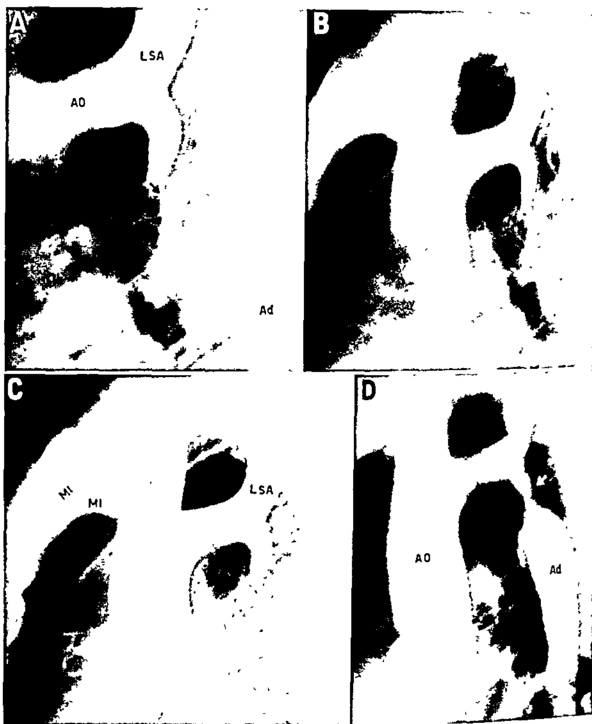


Fig. 561.—Coarctation of the aorta (type II) Man, aged 31 (R.A. 210114) *A–C*, before opera-
[illegible]). Stricture, almost 1 cm long, has
[illegible] dilatation. Left subclavian artery
[illegible] on. Narrow prestenotic segment is
no longer displaced forward (arrow) Considerable reduction in collateral circulation despite relatively narrow anastomosis *Ad*, descending aorta, *AO*, ascending aorta and aortic arch; *LSA*, left subclavian artery, *MI*, internal mammary arteries



Fig 562 —High-lying coarctation of the aorta

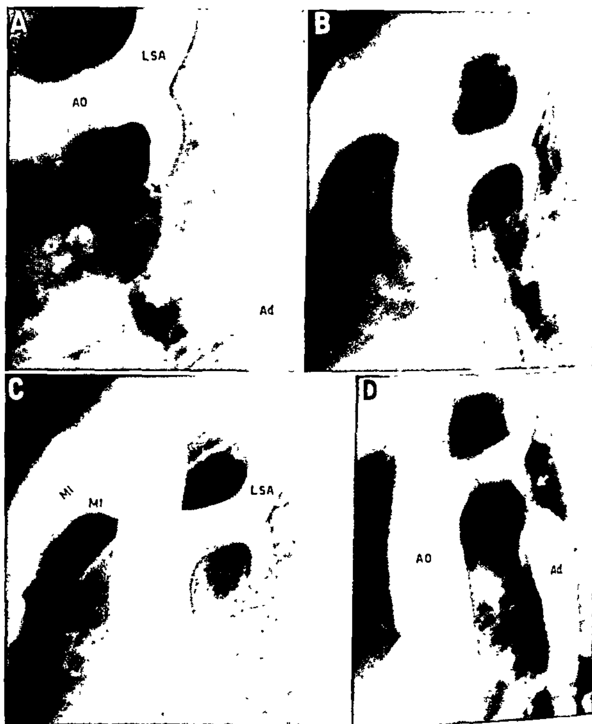


Fig. 561.—[illegible] *A–C*, before opera- [illegible] ost 1 cm long, has [illegible]

[illegible] relatively narrow anastomosis *Ad*, descending aorta, *AO*, ascending aorta and aortic arch, [illegible] left subclavian artery, *MI*, internal mammary arteries

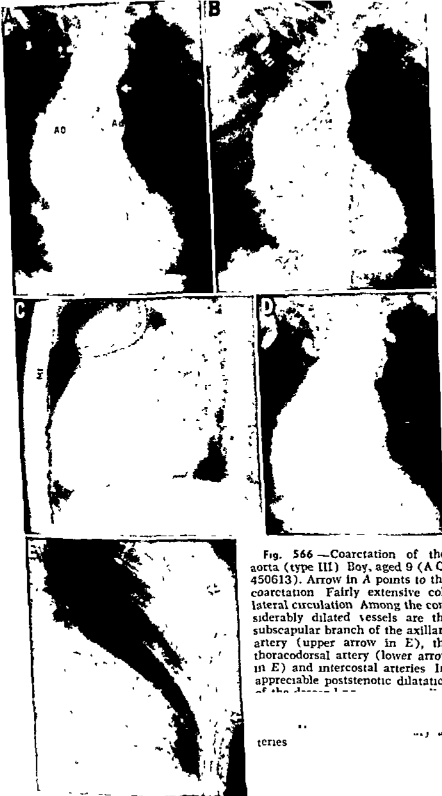


Fig. 566.—Coarctation of the aorta (type III) Boy, aged 9 (A C. 450613). Arrow in *A* points to the coarctation Fairly extensive collateral circulation Among the considerably dilated vessels are the subscapular branch of the axillary artery (upper arrow in *E*), the thoracodorsal artery (lower arrow in *E*) and intercostal arteries Inappreciable poststenotic dilatation of the descend...

teries



Fig. 563 (*left*).—Membranous coarctation of aorta with central orifice about 1 mm wide Child, aged 1 month Arrow points to the orifice. No poststenotic dilatation of the descending aorta. *Ad*, descending aorta, *AO*, aortic arch.

Fig. 564 (*right*).—Membranous coarctation of aorta with large eccentric orifice.



Fig. 565.—Coarctation of the aorta slightly over 1 cm long, with central orifice 2 mm in diameter. Upper arrow in *B* points to orifice In *A*, probe is in the coarctation. Arrow in *A* and lower arrow in *B* point to a short patent ductus, about 1 mm wide. *Ad*, descending aorta, *AO*, aortic arch, *PA*, pulmonary artery

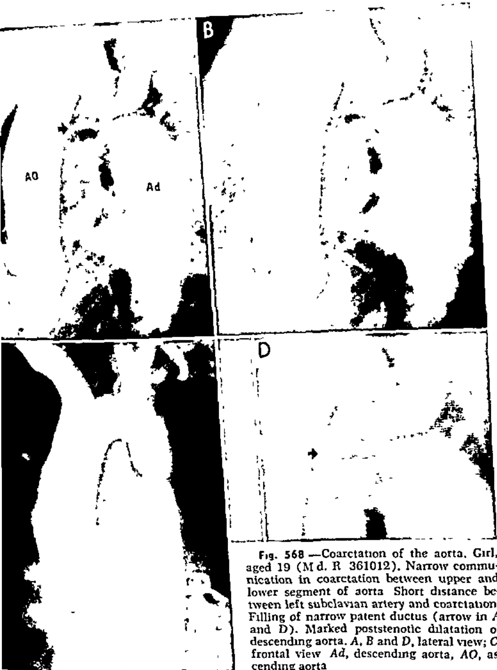


Fig. 568.—Coarctation of the aorta. Girl, aged 19 (Md. R 361012). Narrow communication in coarctation between upper and lower segment of aorta Short distance between left subclavian artery and coarctation. Filling of narrow patent ductus (arrow in *A* and *D*). Marked poststenotic dilatation of descending aorta. *A*, *B* and *D*, lateral view; *C*, frontal view *Ad*, descending aorta, *AO*, ascending aorta

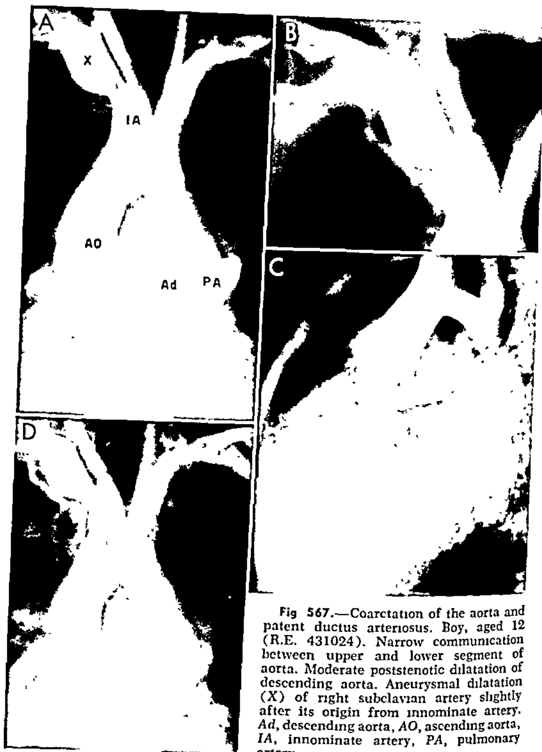


Fig 567.—Coarctation of the aorta and patent ductus arteriosus. Boy, aged 12 (R.E. 431024). Narrow communication between upper and lower segment of aorta. Moderate poststenotic dilatation of descending aorta. Aneurysmal dilatation (*X*) of right subclavian artery slightly after its origin from innominate artery. *Ad*, descending aorta, *AO*, ascending aorta, *IA*, innominate artery, *PA*, pulmonary artery.

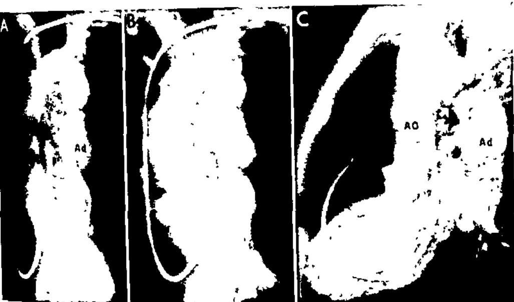


Fig 570 —Coarctation of the aorta. Boy, aged 9 (B E 451228) Contrast medium injected into main trunk of pulmonary artery Narrow communication between two segments of aorta Fairly marked poststenotic dilatation of descending aorta *Ad*, descending aorta; *AO*, ascending aorta



Fig 571 —Coarctation of the aorta Girl, aged 14 (C M. 421022) Fairly well-developed collateral circuit Arrows point to collateral arterial trunks



Fig. 569.—Coarctation of the aorta Ma aged 19 (S-E.P. 350521). Typical forwa and medial displacement of constricted se ment (arrows). Fairly pronounced pos stenotic dilatation of descending aorta (*Ad* Lumen of coarctation (arrow in *A*) about mm in diameter *AO*, ascending aorta.

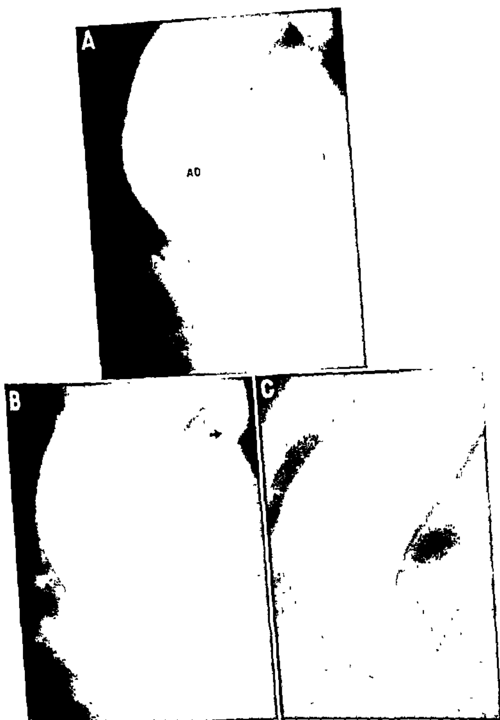


Fig 574 —Coarctation of the aorta Boy, aged 4 (L P 490212) Arrow in *B* points to the coarctation Only two sinuses of Valsalva are visible *A* and *B*, frontal projection *A*, diastole, *B*, systole *AO*, aorta



Fig. 572.—Coarctation of the aorta Man, aged 26 (K.M. 270313). Extensive collateral circulation; dilated collateral arterial trunks *CAT*, collateral arterial trunks, *MI*, internal mammary arteries

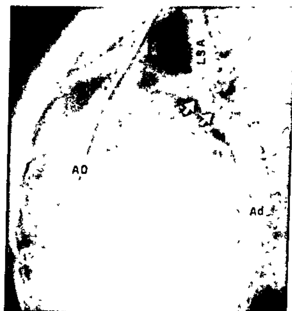


Fig. 573.—Coarctation of the aorta Girl, aged 8 (M L. 431114). Ascending aorta is narrow, no poststenotic dilatation of descending aorta, which is displaced dorsally by the enlarged left [illegible] the origin of the left sub-
descend-

tated into the left ventricle (Fig. 576). A similar but extremely small leakage of contrast medium was seen in two other cases, but aortic incompetence could not be established clinically.

Figure 577 presents both an intercostal aneurysm below the coarctation observed preoperatively and an aneurysm due to rupture of a suture, both of them were identified on thoracic aortography.

In one case, an incidental finding on angiocardiography was an intracardiac de-

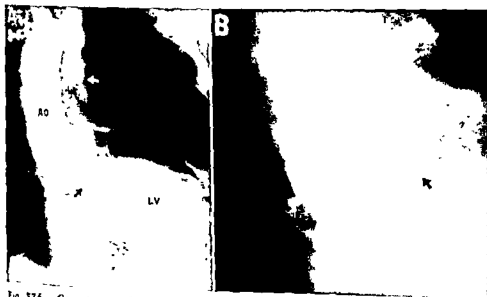


Fig 576 — Coarctat[illegible]
Wh[illegible]
bor[illegible]
in [illegible]

formity interpreted as *herniation of the* ventricular septum (Fig. 64, p. 63).

Very mild coarctation *of the aorta with* no clinical manifestations *was detected* incidentally on angiocardiography in one case of atrial septal defect, in one of mitral incompetence, and in one of interatrial communication through the coronary sinus (Fig 445, p 487) The constriction produced inappreciable deformity of the lumen and caused no displacement of the vessel In one case the coarctation was situated in the abdominal aorta, at the origin of the superior mesenteric and renal arteries

B. COARCTATION OF THE AORTA WITH FETAL TYPE OF CIRCULATION THROUGH THE PATENT DUCTUS

This malformation consists of marked coarctation proximal to a wide patent ductus and poorly developed collateral circulation When there is total aplasia of the aortic arch in combination with an extremely wide ductus, the systemic circulation is divided into two separate circuits. The upper part of the body receives blood from the left ventricle, whereas the lower part is supplied by the right ventricle.

A malformation of this nature is compatible with practically normal circulation during *fetal life* At birth, a sudden, dramatic change occurs. The right ventricle must then supply the pulmonary circulation and, *in addition*, the descending aorta The level *of* the systemic pressure must be maintained by the pulmonary artery. When respiration starts, the resistance in the pulmonary circulation falls. Even if it does not fall *as much* as normally, the pulmonary flow must be very large *to raise* the pressure to the level of the systemic pressure. More-

strated on angiocardiographic examination in only four of them (Figs. 567 and 568). This is in agreement with the observations of Jönsson *et al.* (370) that when the ductus is narrow and the shunt small, demonstration of the ductus is hampered by overlapping collaterals. In many of the aforementioned cases, the contrast medium was injected into the pulmonary artery, which further decreased the possibilities of depicting the ductus. In an additional few of our cases, the contrast medium passed into the pulmonary vessels although the ductus could not be visualized.

In one case a patent ductus was depicted by injecting the contrast medium into it, this resulted in simultaneous depiction of the lower border of the coarctation (Fi 575).

In two cases, there were clinical groun for suspecting associated aortic incomp tence, it was confirmed by thoracic aorto raphy. The contrast medium passed fro the aorta through the orifice and regurg

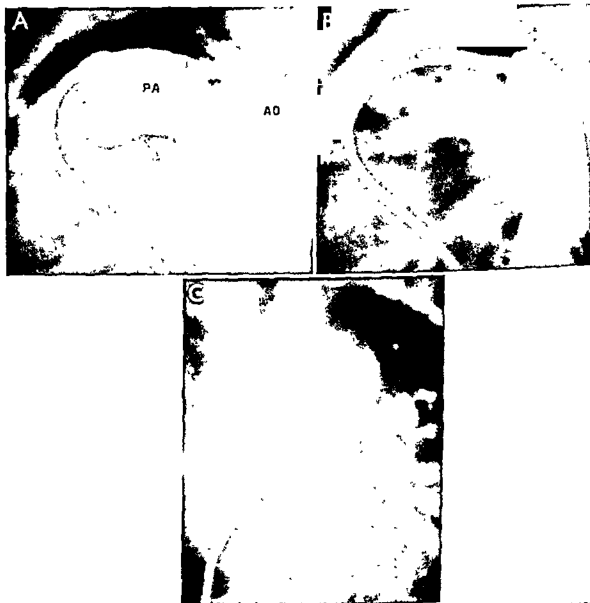


Fig. 575.—Coarctation of the aorta and patent ductus arteriosus. Girl, aged 3 (A.J. 530401 Injection of contrast medium into patent ductus, which lies directly below the subtotal coarct tion Left to right shunt through ductus. *AO*, ascending aorta *PA*, pulmonary artery.

over, the arterial oxygen saturation is low in the lower part of the body. Thus, a highly pathologic circulation is started abruptly and places an enormous load on the heart, mainly on the right ventricle. The blood flow through the left ventricle is also increased in proportion to the increase in the pulmonary flow, but this is insignificant in comparison with the overloading of the right ventricle. Such patients die during the first days of life, presenting a picture of right ventricular failure. If the resistance in the pulmonary vascular bed remains high postnatally, the pulmonary flow is small. The stroke volume of the left ventricle will be small, but sufficient to maintain a normal pressure and flow in the upper part of the body, including the coronary circulation. The stroke volume of the right ventricle will also be small. The load on the heart thus decreases, but the arterial oxygen saturation in the lower part of the body falls concurrently. In such circumstances, very high resistance in the pulmonary [illegible]

[illegible] of the body plays an essential role in regulating the circulation under these exceptional pathologic conditions. Cardiac catheterization has never been performed on such newborn patients. Consequently, the hemodynamic course outlined here is theoretical only, but it is in agreement with the clinical features and the findings at autopsy

Calodney and Carson (129) made a comprehensive study of coarctation of the aorta in early infancy. They found that individuals with a wide ductus have the greatest degree of right-sided dilatation and that "they may survive for several days." They also stated "Those patients with the greatest hypert[illegible]

[illegible]rowest diameter of the ductus, and (3) they had the highest blood pressure." The cases described by Bahn *et al* (33) belonged to this type

The early death has been ascribed to the severe intracardiac malformations which are so often present in these cases (650). This observation does not agree with our experience. Our series comprises four essentially uncomplicated cases and three with intracardiac malformations. The clinical features differed in the two groups. Three of the infants in the first group died during the neonatal period, they presented a picture of right ventricular failure. They were not examined at the cardiologic clinic, but were admitted to the general pediatric department. Autopsy was performed, but the specimens were not kept. The course in these three cases was as follows.

Boy (G L 520927)—Birth weight was 3,700 Gm. Since his first day of life, his mother had noticed that he tired easily and turned gray when he was at the breast. Respiration had always been rapid. At 13 days of age he suddenly became unconscious, he was admitted to hospital in a moribund condition and died 10 hours later. He had an ashen-gray color with marked cyanosis of the lips, considerable edema, and marked dyspnea. Both the radial and the femoral artery pulse were impalpable. The cardiac findings were characterized by a faint systolic murmur and a greatly accentuated second sound over the pulmonary area. A few râles were audible over the lungs. The liver was enlarged. The ECG showed pronounced right axis deviation, only the standard leads were recorded. Autopsy revealed severe coarctation proximal to a patent ductus arteriosus which continued directly into the descending aorta. No intracardiac malformations were present

Boy (T E 521107)—Birth weight was 3,410 Gm. At the age of 6 days he suddenly became cyanotic and dyspneic. When admitted to the pediatric department he was found to have marked dyspnea, slight generalized cyanosis and slight scrotal *edema*. A harsh systolic murmur was heard, but no accentuation of the heart sounds. The veins were dilated. The femoral pulse could not be palpated. The liver was enlarged. Nothing abnormal was heard on auscultation of the lungs. He died two hours later. Autopsy showed enlargement of the right ventricle and severe coarctation proximal to a patent ductus arteriosus [illegible]

A [illegible]

tr[illegible]

[illegible] (T G 540314)—At 1 day of age, she was admitted to the surgical clinic because of atresia of the esophagus, for which she underwent operation. During the first few days



Fig. 577.—Coarctation of the aorta (type I) Girl, aged 16 (B T. 370611) *A* and *B*, before operation Extensive collateral circulation, slight poststenotic dilatation of descending aorta, aneurysmal dilatation of an intercostal artery immediately below the coarctation (arrow in *B*), verified at operation. *C–E*, after operation. A slight constriction at the level of the anastomosis Small postoperative aneurysm at the level of the suture line on the anterior aspect of the vessel (arrow in *D* and *E*). *C* and *D*, frontal projection, *E*, lateral projection *Ad*, descending aorta, *AO*, ascending aorta and aortic arch, *LSA*, left subclavian artery

over, the arterial oxygen saturation is low in the lower part of the body. Thus, a highly pathologic circulation is started abruptly and places an enormous load on the heart, mainly on the right ventricle. The blood flow through the left ventricle is also increased in proportion to the increase in the pulmonary flow, but this is insignificant in comparison with the overloading of the right ventricle. Such patients die during the first days of life, presenting a picture of right ventricular failure If the resistance in the pulmonary vascular bed remains high postnatally, the pulmonary flow is small. The stroke volume of the left ventricle will be small, but sufficient to maintain a normal pressure and flow in the upper part of the body, including the coronary circulation The stroke volume of the right ventricle will also be small. The load on the heart thus decreases, but the arterial oxygen saturation in the lower part of the body falls concurrently. In such circumstances, very high resistance in the pulmonary circulation should be most advantageous for the heart. It must be assumed that a mechanism ...

... pathologic conditions. Cardiac catheterization has never been performed on such newborn patients. Consequently, the hemodynamic course outlined here is theoretical only, but it is in agreement with the clinical features and the findings at autopsy

Calodney and Carson (129) made a comprehensive study of coarctation of the aorta in early infancy. They found that individuals with a wide ductus have the greatest degree of right-sided dilatation and that they may survive for several days " They also stated "Those patients with the greatest hypert... ...y had the highest blood pressure" The cases described by Bahn *et al* (33) belonged to this type

The early death has been ascribed to the severe intracardiac malformations which are so often present in these cases (650). This observation does not agree with our experience. Our series comprises four essentially uncomplicated cases and three with intracardiac malformations. The clinical features differed in the two groups. Three of the infants in the first group died during the neonatal period, they presented a picture of right ventricular failure. They were not examined at the cardiologic clinic, but were admitted to the general pediatric department Autopsy was performed, but the specimens were not kept. The course in these three cases was as follows.

Boy (G L. 520927).—Birth weight was 3,700 Gm Since his first day of life, his mother had noticed that he tired easily and turned gray when he was at the breast. Respiration had always been rapid. At 13 days of age he suddenly became unconscious, he was admitted to hospital in a moribund condition and died 10 hours later. He had an ashen-gray color with marked cyanosis of the lips, considerable edema, and marked dyspnea Both the radial and the femoral artery pulse were impalpable The cardiac findings were characterized by a faint systolic murmur and a greatly accentuated second sound over the pulmonary area A few râles were audible over the lungs The liver was enlarged The ECG showed pronounced right axis deviation, only the standard leads were recorded Autopsy revealed severe coarctation proximal to a patent ductus arteriosus which continued directly into the descending aorta No intracardiac malformations were present.

Boy (T E 521107)—Birth weight was 3,410 Gm At the age of 6 days he suddenly became cyanotic and dyspneic When admitted to the pediatric department he was found to have marked dyspnea, slight generalized cyanosis and slight scrotal edema A harsh systolic murmur was heard, but no accentuation of the heart sounds The veins were dilated The femoral pulse could not be palpated The liver was enlarged Nothing abnormal was heard on auscultation of the lungs He died two hours later Autopsy showed enlargement of the right ventricle and severe coarctation proximal to a patent ductus arteriosus which continued directly into the descending aorta Apart from a small atrial septal defect, no intracardiac malformations were found.

Girl (I G 540314)—At 1 day of age, she was admitted to the surgical clinic because of atresia of the esophagus, for which she underwent operation During the first few days

after operation her condition was satisfactory, but when she was 9 days old a sudden exacerbation took place, with generalized cyanosis and severe dyspnea. Death occurred 24 hours later. No murmur had been audible. Autopsy disclosed severe coarctation proximal to an extremely wide ductus arteriosus which continued directly into the descending aorta. The esophageal anastomosis had healed satisfactorily, and the cause of death was considered to be heart failure.

Roentgenologic examination of these three newborn infants, none of whom had had never shown any signs of heart failure. Her physical development was retarded, she had not learned to walk Cyanosis was only periodic. Physical examination showed an accentuated, pure second sound over the pulmonary area but no murmur The blood pressure, determined with the flush method (284), was 150 mm Hg in the arms and 90 mm in the legs The ECG showed marked right ventricular hypertrophy.

Roentgenologic examination showed only moderate enlargement of the heart, with marked right ventricular hypertrophy The pulmonary vessels had the same appearance as

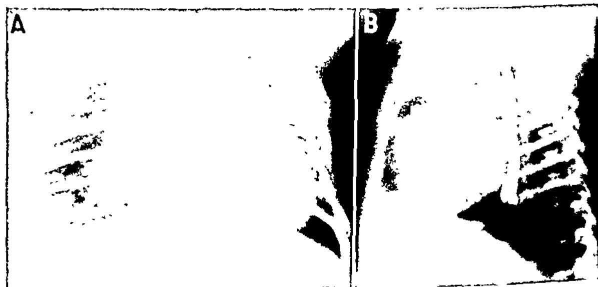


Fig. 578.—Coarctation of the aorta with fetal type of circulation through the ductus Boy, aged 1 week (T E. 521107). Definite enlargement of heart, without normal postnatal reduction in volume, increased vascularity of lungs, left atrium of normal size, the thymus overlaps the great vessels

any intracardiac malformation, showed a definite increase in the heart volume, which seemed to be due to dilatation of the right atrium and right ventricle. Vascularity of the lungs was augmented (Fig. 578). The anatomic details of the aorta and the pulmonary artery could not be identified, but they are hard to evaluate at this age, even in normal cases. The appearance is in agreement with the few descriptions found in the literature (326, 476, 650), but it is far from characteristic, and it would scarcely be possible to make an exact roentgenologic diagnosis on the basis of these findings alone.

The features differed in the fourth case.

GIRL, AGED 1½ YEARS (A.D 541112) —She in high pulmonary vascular resistance. On angiocardiography with injection into the right ventricle, the contrast medium passed from the pulmonary artery into a wide patent ductus arteriosus which continued directly into the ascending aorta (Fig. 579). The aortic arch was atresic slightly proximal to the ductus arteriosus, and no collaterals could be demonstrated

The following values were obtained on *cardiac catheterization:*

	O2 Content Vol %	Pressure mm Hg
SVC	11 9	
RA	9 8	1
RV	9.3	90/4
PA	8 8	71/49
Descending aorta	9 2–48% sat	71/49
O_2 cap	19 0	

Since the oxygen content was the same in samples from the pulmonary artery and

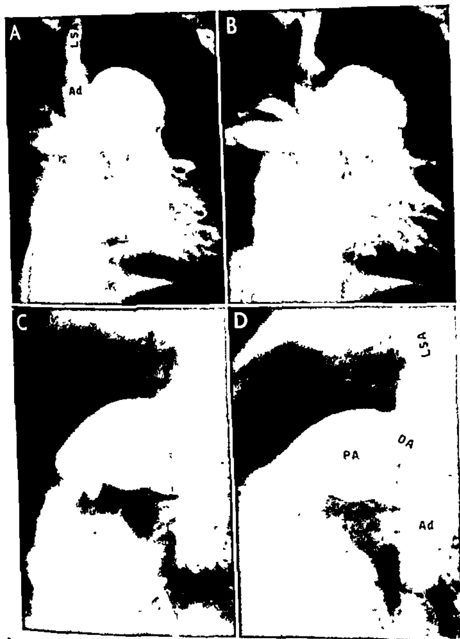


Fig 579 —Coarctation of the aorta with fetal ... 541112). Cor... through a w... subclavian a...

descending aorta, there could have been no passage of oxygenated blood from the ascending to the descending aorta. This observation agrees with the angiocardiographic findings. If it is assumed that the pulmonary venous blood had an oxygen saturation of 97 per cent, the arteriovenous oxygen difference was 10 volumes per cent, and the pulmonary flow was therefore very small As mentioned above, the roentgenograms indicated the presence of

Boy, aged 2 months (R H. 521220) —Heart disease was diagnosed when he was 1 month old. He then became cyanotic during feedings and when he cried, but he had no dyspnea or edema. He gained poorly in weight. The cardiac findings were characterized by a greatly accentuated second sound over the pulmonary area and a systolic murmur over the second right interspace. The femoral pulse was palpable. At 20 months of age he exhibited no cyanosis at rest but only on exertion, it was not more marked in the lower part of the body. The ECG showed right ventricular hypertrophy and abnormally tall P waves in V_1.

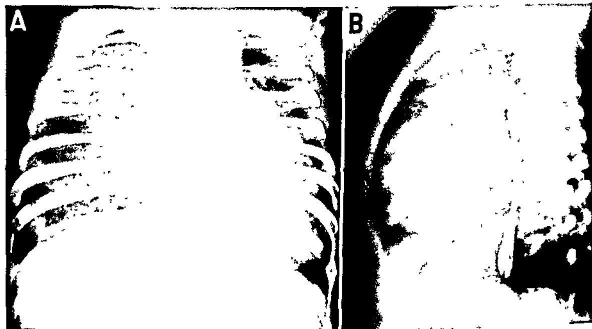


Fig. 580.—Coarctation of the aorta with fetal type of circulation, transposition of aorta and over-riding of pulmonary artery (Taussig-Bing complex). Boy, aged 7 weeks (R H. 521220), see Figure 582 Greatly increased heart volume with enlargement of right atrium and ventricle, dilatation of pulmonary artery, greatly increased vascularity of lungs, left atrium of normal size, narrow aorta

high pulmonary vascular resistance, thus distinguishing this case from the other three. The high resistance seems to account for the patient's survival

None of the patients *with an intracardiac malformation* showed any symptoms during the neonatal period. In one case there were a ventricular septal defect and incomplete transposition of the great vessels (Taussig-Bing complex), and in the other two there was a moderately large ventricular septal defect Both cardiac catheterization and angiocardiography were performed.

Boy, aged 9 years (L A 420531) —Heart disease was detected, owing to a murmur, on a routine examination during his first year of life He started to walk at 1 year of age. There after he easily became tired on exertion and s[illegible] was m[illegible] ment [illegible] ity N[illegible] findings were characterized by a greatly accentuated second sound and a loud, harsh systolic murmur over the pulmonary area The blood pressure was 185/95 mm Hg in the arms and 150/90 mm Hg in the legs The ECG showed right ventricular hypertrophy

Girl, aged 8 months (E T 551005).—At [illegible] months of age a murmur was heard, and congenital heart disease diagnosed She gained

poorly in weight There was no cyanosis or dyspnea The blood pressure (flush method) was 140 mm Hg in the arms and 105 mm in the legs A pansystolic murmur was audible over the apex, and the second sound was accentuated over the pulmonary area. The ECG showed marked right ventricular hypertrophy, but also signs of left ventricular hypertrophy

When coarctation of the aorta of this type is combined with an intracardiac shunt, *the roentgenologic appearance* is dominated by the increased blood flow in the pulmonary circulation. This has also been pointed out by Seaman and Goldring (586)

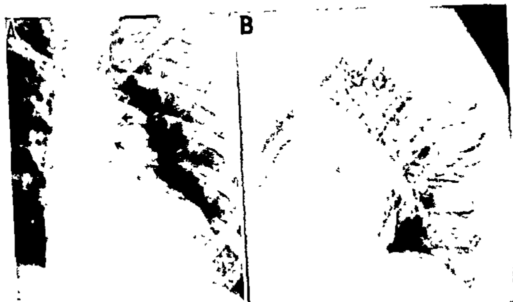


Fig 581 ... fetal type of circulation through the ductus Boy ...

In one infant (R H 521220), the right atrium, right ventricle and presumably the left ventricle as well were enlarged (Fig 580) The pulmonary artery and its branches were dilated, as in the presence of an increased blood flow in the pulmonary circulation No exact evaluation of the aorta could be made In the other infant (E.T. 551005), the whole heart was enlarged, not least the right and left atria The lungs exhibited increased vascularity as a sign of a left to right shunt. The anomalous origin of the subclavian artery produced a typical impression in the esophagus.

In the 9-year-old boy (L A. 420531), there were definite signs of a left to right shunt (Fig 581), consisting of considerable dilatation of the pulmonary artery and its branches, enlargement of the right ventricle, and suggested dilatation of the left atrium The aortic arch could not be visualized either on the roentgenogram or on fluoroscopy The ascending aorta was narrow The descending aorta was clearly visible below the pulmonary artery In the lateral projection, a vessel outline appeared which, on comparison with the angiogram, could be identified as the superior border of the ductus arteriosus

The findings on *cardiac catheterization* are recorded in Table 18 In these cases, the characteristic feature of the hemodynamics is that the pulmonary artery and the de-

scending aorta have the same pressure and practically the same oxygen content. In one case (E.T. 551005) there was, however, a small blood flow through the coarctation, and the oxygen content of the descending aorta was slightly higher than that of the pulmonary artery. In both cases with a ventricular septal defect, the oxygen saturation of the blood in the descending aorta was only slightly decreased, owing to the left to right interventricular shunt. It was, however, decreased to such a degree that samples from the inferior vena cava had a definitely lower oxygen content than samples from the superior vena cava. The diagnosis could be established on the basis of these findings.

Patient R H. (521220) was only 2 months old at the time of examination. At this age, the oxygen saturation may vary appreciably from one minute to the next, as in this case The samples from the superior and inferior venae cavae were taken in immediate sequence at the beginning of the examination. A considerably lower oxygen content was found in the former, which is understandable in view of the fact that the upper part of the body was supplied by the right ventricle In this complicated case, the anatomic diagnosis could be made only with the help of the angiocardiographic findings.

The *angiocardiographic examination* in this case (Fig 582) was made by injection of the contrast medium into the right ventricle The aorta was transposed and the pulmonary artery over-rode a wide ventricular septal defect, i e , Taussig-Bing complex. The distal segment of the aortic arch was aplastic. The pulmonary artery communicated with the descending aorta through the ductus arteriosus, which had the same width as these vessels The internal mammary and intercostal arteries presented the normal anatomic appearance

The other infant (E T , 551005) was examined at 8 months of age. On angiocardiography with injection of contrast medium into the right ventricle, a typical ventricular septal defect could be demonstrated at the site of the membranous part

TABLE 18 —MOST IMPORTANT FINDINGS ON CARDIAC CATHETERIZATION IN 3 CASES OF COARCTATION OF THE AORTA WITH FETAL TYPE OF CIRCULATION THROUGH A PATENT DUCTUS AND AN INTRACARDIAC MALFORMATION*

Case	O_2 Content, vol %										O_2 Cap	Pressure, mm Hg							Assoc Malform
	SVC	IVC	RA	RV	PA	Aorta		LA	PV	LV		RA	RV	PA	Aorta		LA	LV	
						Asc	Desc								Asc	Desc			
L A 420531 (9 yr.)	17.4	15.1	16.5	19.8	18.8	23.5	19.0	—	—	—	23.5	2	120/8	110/50	180/90	85/50	—	—	VSD
R H 521220 (2 mo)	6.2	9.9	8.5	15.3	13.6	—	14.4	18.0	17.5	17.1	19.4	2	77/4	75/40	—	80/32	6	60/4	Taussig-Bing complex
E T 551005 (9 mo.)	10.1	8.4	10.3	12.0	12.0	—	13.5	—	—	—	15.6	2	85/10	81/36	—	71/46	—	—	VSD

*For abbreviations see Table 1, p 119

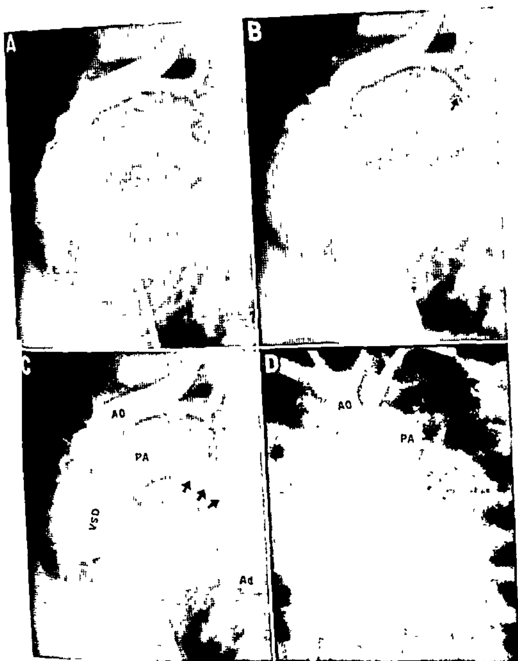


Fig 382.—Coarctation of the aorta with fetal type of circulation, transposition of aorta, and overriding of pulmonary

PA, pulmonary artery, VSD, ventricular septal defect.

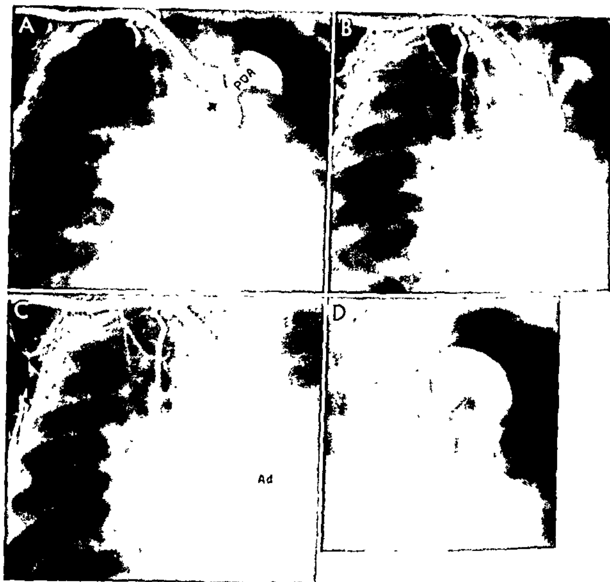


Fig. 583.—Coarctation of the aorta with fetal type of circulation and ventricular septal defect. Girl, aged 8 months (E T. 551005) On injection of tery, opacification is obtained in diastole both of the ao (arrow in *A* and *G*) and of descending aorta (*Ad*) below tus (*PDA*) and part of the pulmonary artery (*PA*) In ductus in the opposite direction, and the contrast medium therefore flows into the descending aorta (*C* and *E*) (*continued*)

of the septum, below the crista superventricularis The pulmonary artery, ductus arteriosus, and descending aorta continued into each other without any definite borderline. At complementary aortography, performed by retrograde injection into the right brachial artery, it could be established that a communication was present in the coarctation, which was a membranous type (Fig. 583).

In the 10-year-old boy (L.A. 420531) we performed both thoracic aortography and, at a later session, injection of the contrast medium into the pulmonary artery.

Thoracic aortography (Fig 584, *A–D*) showed that the distal segment of the aortic arch was aplastic and the ascending aorta narrow Only scanty collaterals could be identified. Slight filling of the descending aorta gradually took place through its communications. In this form of coarctation it is advisable to use a smaller quantity of

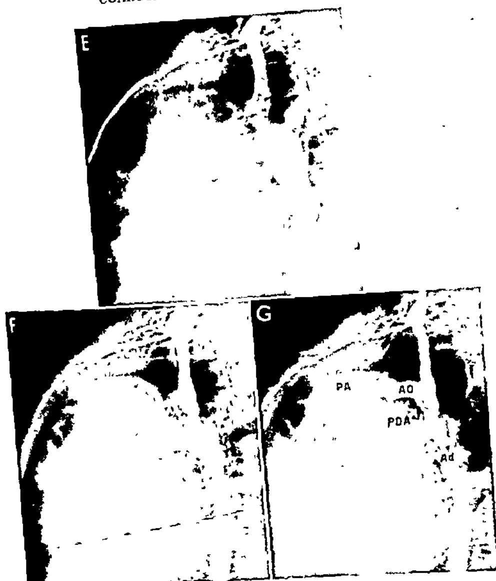


Fig 583 (*cont.*)

contrast medium than usual for thoracic aortography, in order to avoid an overdose to the cerebral vessels

When the contrast medium was injected into the pulmonary artery (Fig 584, *E–F*), a direct communication was visible between the pulmonary artery and the descending aorta As in the previous case, this communication consisted of a patent ductus of the same width as the vessels in question. The coronary vessels to the right side of the heart were definitely wider than the corresponding vessels to the left ventricle, presumably as a manifestation of the increased burden on the right ventricle. The atresic segment of the aorta was about 5 cm long.

Thus in all our cases a characteristic feature was that the pulmonary artery communicated directly with the descending aorta through the ductus arteriosus, without causing any displacement of the aorta. The ventral displacement of the aorta at the level of the constriction, which is typical feature in all usual forms of co

Fig. 584 —Coarctation of the aorta with fetal type of circulation Boy, aged 9 (L A 420531) ... artery to mouth of the ... artery is ... le (*A–D*) ... ce to the

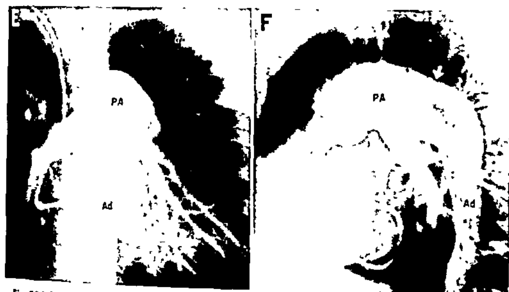


Fig 584 (*cont.*) —E–F, injection of contrast medium into the dilated pulmonary artery. The wide patent ductus continues in an even arch directly into the descending aorta (arrow in *F*). *Ad*, descending aorta, *PA*, pulmonary artery.

arctation of the aorta, was thus lacking in these cases

From the *clinical and hemodynamic viewpoints*, these cases have a great similarity to a patent ductus combined with high resistance in the pulmonary circulation and a reversed shunt only, *these cases* being unsuitable for operation. It is true that no severe symptoms were present in case L A 420531, but the prognosis was regarded as poor in view of the hypertension and the great overburdening of the right ventricle. It was considered that the hypertension in the upper part of the body could be eliminated by surgical treatment of the vascular anomaly, even if the circulation could not be entirely normalized Operation was therefore performed on this indication. Using a graft, an anastomosis was made between the ascending and descending aorta. The patent ductus was then closed. The course was satisfactory during the first days after operation, but heart failure developed after a week, and the patient died.

It thus appears as though cases of coarctation of the aorta with a fetal type of circulation through the patent ductus have a better prognosis if an intracardiac malformation is present, permitting some oxygenated blood to pass into the descending aorta The patients may, however, die early (40, 597). On the other hand, children with no associated intracardiac malformation have been known to survive for some time (222, 638, 653), as in *one of* our cases.

19 Vascular Ring

VASCULAR RING as a clinical concept denotes those anomalies of the aortic arch system which cause symptoms of tracheal and esophageal compression. In contradistinction to other malformations of this system (coarctation of the aorta and patent ductus arteriosus), vascular ring gives rise to no cardiac signs or symptoms. A patent ductus often forms an essential part of the vascular ring, but it may be obliterated.

The anatomy of the vascular ring varies Edwards (216) has classified the different types according to a basic pattern of the aortic arch system, with a double aortic arch and double ductus arteriosus. The various types may develop from obliteration or aplasia of certain parts of this vascular system (Fig. 64, p. 63). There is sometimes a functioning double aortic arch, but some parts of the ring may lack a lumen or be entirely resorbed. In the latter event, there is no ring from the anatomic viewpoint, but the anomaly may still cause symptoms of compression (Fig 585).

From the clinical standpoint, the only factor of importance is the degree of constriction. Some forms of the anomaly are asymptomatic. This applies particularly to the most common type a left-sided aortic arch and left-sided descending aorta, the right subclavian artery is given off by the descending aorta and passes behind the esophagus, or between the trachea and the esophagus. This type occurs in 0 5 per cent of all individuals.

If the ring is narrow, severe symptoms appear as early as the neonatal period. According to Taussig (650), the compression decreases as the child grows, and the malformation therefore causes no symptoms in older children and adults. In our opinion, the most severe forms are incompatible with life, and the patients die in infancy of asphyxia or pneumonia.

We have three cases with typical symptoms in *our series;* a brief account of them is given here to illustrate the clinical features

GIRL, AGED 5 DAYS (C Z. 500717) —Admitted to our hospital because of stridor, she exhibited marked air hunger and deep respiratory retractions. Feeding presented great difficulties, about 1½ hours were required for each feeding, since there was constant regurgitation. She gained weight poorly. Her condition deteriorated, and operation was therefore performed [illegible] confirm[illegible] been re[illegible] (229).

BOY, AGED 18 MONTHS (J.G 500630).—Stridor had been present since birth, *but there* was no appreciable air hunger at rest and respiration was completely normal during sleep He contracted repeated respiratory tract infections but had no dysphagia He was brought to our outpatient department for asthmatic children The diagnosis of vascular ring was verified at operation

BOY, AGED 1 MONTH (H K 540120) —Since birth he had exhibited both inspiratory and expiratory stridor, with retraction in the epigastrium He had not suffered from vomiting and gain in weight was satisfactory. The diagnosis was verified at operation



Fig 585 —Vascular ring Boy, aged 3 months (T N 530527) Compression of trachea and esophagus slightly above the bifurcation

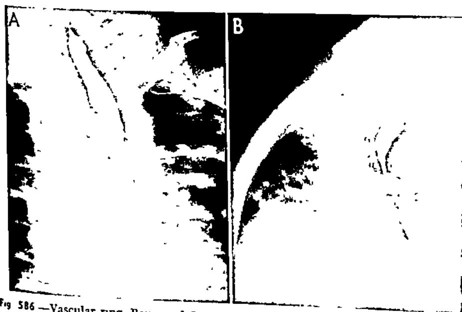


Fig 586 —Vascular ring Boy, aged 6 months (M Z 541103). Compression of the trachea and esophagus is caused by the right aortic arch which runs behind the esophagus and by a fairly wide patent ductus arteriosus and left subclavian artery

The presence of a vascular ring should be suspected in newborns with stridor associated with air hunger or dysphagia, and in cases in which stridor persists for any length of time. Owing to pressure on the recurrent laryngeal nerves, the voice is often hoarse. The diagnosis is established by roentgenologic examination, at which compression of both the trachea and the esophagus can be demonstrated (505) (Figs. 585 and 586). Thoracic aortography seems to be superfluous and often useless. Part of the ring may consist of fibrous strands which cannot be visualized with this technique. Moreover, the indications for operation are based on the clinical syndrome, not on the anatomic details.

A special form of vascular anomaly, often associated with respiratory distress, is caused by an abnormal course of the left pulmonary artery. It arises to the right of the midline, runs backward and then to the left between trachea and esophagus. The condition was first described in 1954 by Welsh and Munro (684) and by Potts *et al.* (541). Additional cases have been reported by other authors (335, 709). Roentgenologically, this vascular anomaly is disclosed by a characteristic indentation in the anterior wall of the esophagus, just below the level of the aortic arch, and compression of the lower part of the trachea or right main bronchus. The abnormal course of the pulmonary artery can be confirmed on angiocardiography (335) Emphysema may be present bilaterally or in the right lung only.

20 Valvular and Subvalvular Aortic Stenosis

CONGENITAL AORTIC stenosis may be valvular, the cusps then being fused into a dome (Fig 612), or subvalvular (subaortic stenosis) (Fig 593). In the latter type, the stenosis is caused by a fibrous ring in the infundibulum of the left ventricle, immediately beneath the membranous part of the ventricular septum and close to the anterior cusp of the mitral valve, which is sometimes involved The cusps of the aortic valve may be thickened, owing to secondary changes (216, 439) (Fig. 597b).

It does not seem possible at present to distinguish clinically between valvular and subaortic stenosis. Some authors (100, 107, 326) have stated that the physical findings differ significantly in the two types, but others have been unable to verify this statement (137, 386). According to Campbell and Kauntze (137), the presence of calcifications is the most conclusive evidence of valvular stenosis As a rule, however, they are not visible roentgenologically until adult age Moreover, calcifications have also been observed in subaortic stenosis (292)

Many authors are of the opinion that subaortic stenosis is the more common type (650, 713), but others have found a high incidence of valvular stenosis as well (137, 386) This discrepancy may be explained by the difficulty in determining whether valvular stenosis is of congenital or of rheumatic origin Such determination is not always possible even at autopsy. If the stenosis is diagnosed before 3 years of age, it is an argument in favor of a congenital malformation, but does not afford conclusive proof Furthermore, if the heart disease is detected only later in childhood or in adulthood, a rheumatic origin cannot be ruled out, even though there is no history of clinically manifest rheumatic fever or chorea. Calcifications of the valves are common in rheumatic aortic stenosis, but [illegible]

probably responsible for the stenosis (377, 439, 360)

Our series includes 23 children with congenital aortic stenosis. In addition, two patients from other clinics underwent roentgenologic examination only. In three cases, aortic stenosis was combined with aortic incompetence, in one case with coarctation of the aorta and in one case with partial anomalous venous return and coarctation of the aorta. Mild aortic stenosis was also observed in five cases of patent ductus arteriosus (see p 532). In no case was there a history of suspected rheumatic fever or chorea In eight cases the heart disease was, however, diagnosed only after 5 years of age, but in 13 it was diagnosed before the age of 3 years. Subvalvular stenosis was present in two cases in which autopsy was performed. In three other cases the

anatomic diagnosis was established by angiocardiography; valvular stenosis was present in two of them and subvalvular in one.

The sex distribution (19 boys and four girls) showed, as in other series, a large preponderance of males.

CLINICAL FEATURES

Physical underdevelopment has been regarded as common in aortic stenosis (107). All of our patients were normally developed (Fig. 587).

Fourteen of our patients were entirely asymptomatic. In the other nine there was increased fatigability (seven cases), dyspnea (four cases), anginal pain (three cases), and attacks of syncope (one case). A 14-year old girl (B.D., 370625) had displayed dyspnea on exertion since her second year of life. During the past two years deterioration had taken place, and she suffered from great fatigue and anginal pain. One year after examination by us, she died suddenly. Autopsy disclosed severe subaortic stenosis (about 2 × 4 mm) 2 to 3 mm below the base of the aortic valve. The posterior cusp was thickened, and wart-like elevations were present on the convexity. The appearance of the other cusps was normal The left ventricle was grossly hypertrophied An 11-year-old boy died suddenly at play. He had had no cardiac symptoms earlier. The ECG was normal, and roentgenologic examination had shown no enlargement of the heart but slight left ventricular hypertrophy. On the phonocardiogram, the appearance of the murmur was that in only moderately severe stenosis. Autopsy showed the presence of subvalvular aortic stenosis

In many cases, congenital aortic stenosis is so mild that it gives rise to no symptoms. Even the long-term prognosis is good in such cases (100, 107, 502) Stenosis may, on the other hand, be so severe that symptoms appear in childhood. Sudden death is not uncommon and may occur at an early age (88, 479, 502). The patient generally has a history of one or more attacks of syncope, but sudden death has been known to occur without any previous cardiac symptoms, as in one of our cases. In some of such cases both heart volume and electrocardiogram have been normal. Bacterial endocarditis is also a common cause of death.

Physical Working Capacity

The physical working capacity may be normal in mild aortic stenosis. By means of a rise in pressure in the left ventricle, a normal stroke volume can be maintained both at rest and during exercise. Evaluation of the physical working capacity is, however, hampered by the fact that such patients should not work under such a heavy load that the pulse rate rises above 150 per minute.

In one of our severest cases, that of a 16-year-old girl, the physical working capacity was estimated to be only 200 kgm per minute. The stroke volume was small even at rest (52 ml).

Physical Signs

Aortic stenosis in children is easily diagnosed on clinical examination, owing to the typical nature of the murmur. A loud, harsh, systolic murmur is heard; it has a maximum over the aortic area and is transmitted to the large vessels of the neck. The intensity of the murmur increases with the degree of stenosis. In mild cases, the murmur is the only physical finding. In severe stenosis, the left ventricle becomes hypertrophied; the cardiac impulse is increased in breadth, heaving and displaced *laterally* Symptoms from the peripheral circulation are also present in the form of low blood pressure and a small pulse amplitude.

According to Brofman and Feil (100), it is possible *to* differentiate clinically between valvular and subaortic stenosis. They state that the latter is associated with a distinct second sound over the *aortic* area, whereas in the former type the second aortic sound is weaker or lacking This difference has also been stressed by Brown

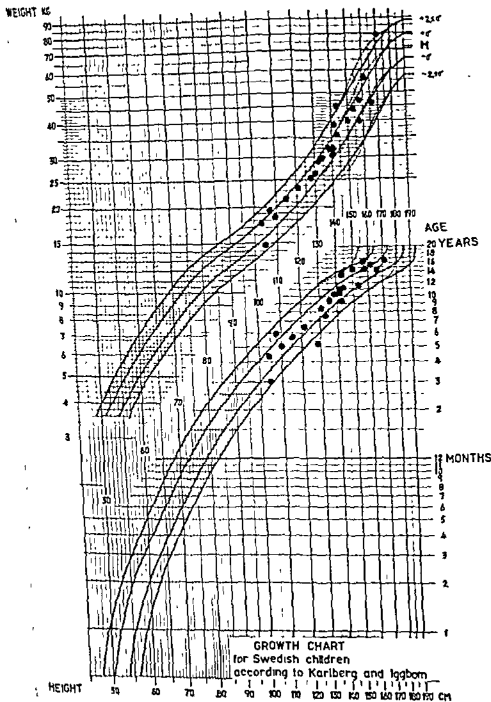


Fig 587 —Physical development in 23 children with valvular or subvalvular aortic stenosis. Both height and weight are normal (Relevant growth chart was kindly placed at our disposal by Drs P Karlberg and S Iggbom [374, 375])

(107). Kiloh (386) has stated, on the contrary, that the second aortic sound is of no diagnostic value. Even in cases with definite valvular stenosis, confirmed at autopsy, the second sound was sometimes entirely normal. A diastolic murmur is considered to be an indication of a valvular lesion (137, 386), but it may also be present in subaortic stenosis when the cusps are deformed (326, 683).

All of our patients presented a characteristic, harsh systolic murmur with a maximum over the second right intercostal space and transmission to the arteries of the neck. In the most severe stenoses, the murmur was so strong that a thrill was palpated over the aortic area and the vessels of the neck. The second aortic sound was normal in all but three cases. All three patients had subaortic stenosis established at operation or autopsy; the second aortic sound could be neither heard nor recorded phonocardiographically.

The phonocardiogram in aortic stenosis has been described by Leatham (414), among others. Characteristically, the systolic murmur is *diamond-shaped*. Such a murmur was recorded in all of our cases. We found a distinct difference between the mild and the severe cases of stenosis. Cases with cardiac symptoms and electrocardiographic and roentgenologic signs of left ventricular hypertrophy exhibited a systolic murmur with extremely high amplitudes. The murmur started immediately after the first sound, reached a maximum in midsystole or somewhat later, and ended only with the second sound. In mild cases (no symptoms, normal heart size, and normal ECG), the amplitude of the murmur was lower, reached a maximum during the first half of systole, and ended 0.04 to 0.06 sec before the second sound. Examples are shown in Figure 588, in which *A* is a phonocardiogram in mild stenosis, *B* in moderately severe stenosis, and *C* and *D* in severe stenosis. An aortic early systolic sound (417) (*claquement aortique protosystolique* (432)), was demonstrable in nine cases; aortic incompetence was present in two of them. This loud, sharply delimited sound of high frequency occurs 0.06 to 0.08 sec after the beginning of the first sound and is recorded best over the apex, where it is not masked by the murmur (Fig. 588, *B*).

In three cases there were coincident aortic stenosis and incompetence, the latter probably being the dominating feature. A characteristic to and fro murmur was audible in every case; its diastolic component was often most distinct over the third left interspace (Fig. 589). According to White (699), no case of isolated congenital aortic incompetence has been described. As a rule, it is combined with some other cardiac or aortic malformation (107). Our series contains two cases in which aortic incompetence was combined with coarctation of the aorta. If there is considerable regurgitation, the stroke volume of the left ventricle is also large, and an early systolic ejection murmur of moderate intensity can be heard without the presence of aortic stenosis.

Low systolic pressure and a small pulse amplitude were present in 12 cases and were most conspicuous when the stenosis was severe. The three patients with coincident aortic stenosis and incompetence had low diastolic pressure as well.

Characteristic changes in the contour of the arterial pulse curve are present in aortic stenosis (100, 303, 579). Piezoelectric recording of the carotid pulse shows a slow rise, a plateau formation with vibrations, and a slow decline. The femoral pulse curve, on the contrary, has a normal contour. Brofman and Feil (100) have stated that it is possible to distinguish between valvular and subvalvular stenosis on the basis of the pulse curve. In valvular stenosis they found the incisura and the dicrotic wave with its following vibrations to be lacking and the fall of the descending limb to be slower. In subaortic stenosis, there was stated to be a more rapid fall, a distinct incisura, and a dicrotic wave followed by vibrations. These conclusions were based on clinical investigation of 10 cases, which were not verified at autopsy. Piezoelectric recordings of the pulse curve were made in about half of our cases, a change in the carotid pulse curve was noted in all of them (Fig. 590). In

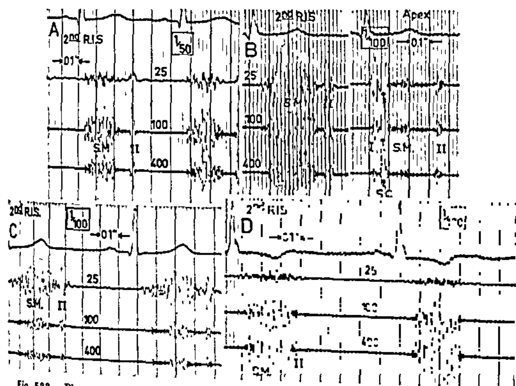


Fig 588 — Phonocardiograms in valvular and subvalvular aortic stenosis *A* mild stenosis
Boy, aged 9 (J A 450509) No ...
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one case confirmed at autopsy—severe subaortic stenosis—there was only a suggestion of an *incisura* and a dicrotic wave

In our opinion, therefore, it is impossible to distinguish between valvular and subaortic stenosis by means of either the auscultatory findings or the pulse tracings. The intensity of the second sound and the shape of the pulse curve seem to depend instead on the degree of stenosis In severe subaortic stenosis, the second aortic sound was diminished, despite the fact that there was only a slight lesion of the valve. A hypothetical explanation is that, in severe subaortic stenosis, closure of the cusps is damped by the blood present in the space

...nosis, when the cusps still retain some flexibility, the occurrence of a sound on their inversion at the beginning of diastole is plausible, since the diastolic pressure in the aorta is *normal*, whereas the pressure

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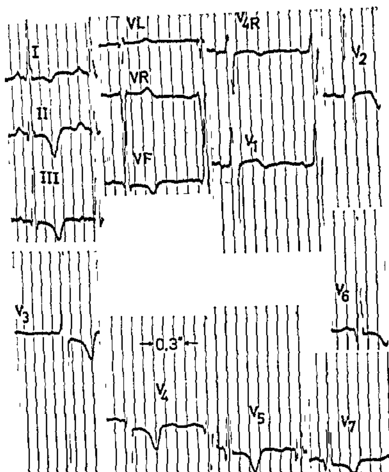


Fig 591.—Electrocardiogram in severe subaortic stenosis Girl, aged 14 (B D 370625) Sudden death one year later Marked left ventricular hypertrophy

behind the cusps—*in the ventricle*—is low The only definite conclusion we can draw from our series is that the *second sound* over the aorta may be lacking in cases of severe subaortic stenosis.

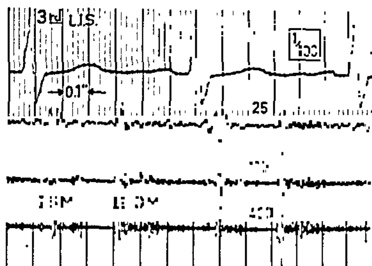
ELECTROCARDIOGRAPHY

Aortic stenosis is frequently so mild, and the extra burden on the left ventricle so slight, that no signs of left ventricular hypertrophy appear on the electrocardiogram In 14 of our cases the ECG was normal, and in the *other* nine there were signs of isolated left ventricular hypertrophy (Fig 591) Among the latter were all three cases of combined aortic stenosis and incompetence Thus, *only* six of the 20 cases of isolated stenosis exhibited electrocardiographic changes. The changes become more marked on exertion (Fig. 592) and may appear in cases with a normal ECG at rest

ROENTGENOLOGIC EXAMINATION

Observations in large series of cases of *congenital* aortic stenosis (107, 137, 207,

The ascending aorta is dilated and its pulsations have an increased amplitude. The aortic arch, on the contrary, is not prominent It may be normal in appearance or narrow The left ventricle is enlarged and



[illegible] stenosis. Boy, aged 6 (S.T. [illegible] degree of amplification, other figures denote standard frequencies of the filters.

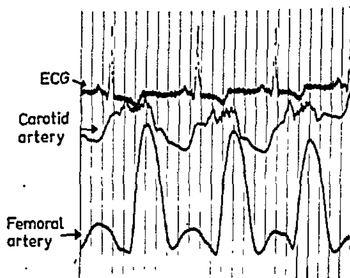


Fig. 590.—Arterial pulse curves in subaortic stenosis Girl, aged 14 (B D. 370625), see Figure 588, D Piezoelectric technique with simultaneous recording Carotid pulse curve is sinus-shaped, with slow ascent and the systolic murmur on the plateau of the tracing Although the femoral pulse curve is slightly damped, its appearance is normal.

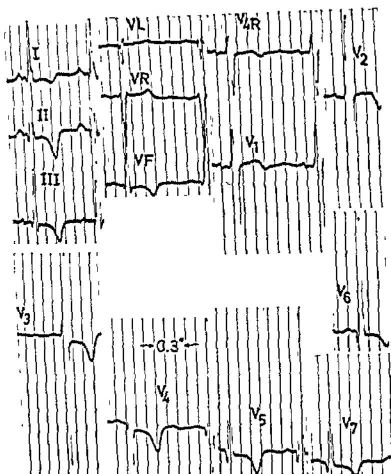


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Electrocardiography

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Roentgenologic Examination

Observations in large series of cases of congenital aortic stenosis (107, 137, 207, 386, 458) have shown that the roentgenologic appearance has the following characteristics

The ascending aorta is dilated and its pulsations have an increased amplitude. The *aortic arch*, on the contrary, is not prominent It may be normal in appearance or narrow The left *ventricle* is enlarged and

exhibits increased curvature and elongation. None of these features is, however, consistently present.

We made a roentgenologic examination in 25 cases of congenital aortic stenosis; all of the patients were under 19 years of age. In four there was coincident aortic incompetence, *which was probably the dominating lesion in three of them* (p. 647). Coarctation of the aorta was present as an associated defect in an additional two cases, in one of them with anomalous pulmonary venous return as well. In one case bulge of the suprabulbar segment of the aortic outline (Figs. 594 and 595). This is usually particularly distinct in the left oblique projection and is also well visualized in the frontal projection. If the aorta can be demonstrated in the lateral view, it is then seen to be closer to the thoracic *wall than normally. In one of our cases* (Fig. 608) angiocardiographic examinations, performed in the frontal and lateral projections and in oblique projections, showed that both dilatation and an abnormal course are responsible for the change

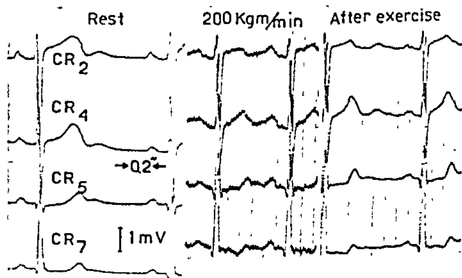


Fig. 592.—Electrocardiogram before, during, and after exercise (200 kg/min) in subaortic stenosis. Boy, aged 13 (K.S. 440512). Pressure in left ventricle 185/15 mm Hg. At rest, only slight depression of S–T interval in CR_{5-7}. During and immediately after exercise, the depression is more marked

of valvular aortic stenosis and coarctation of the aorta, the picture was dominated by a coincident wide patent ductus arteriosus, this case is therefore accounted for in Chapter 16. The angiocardiogram obtained after closure of the ductus, which illustrates the aortic anomalies, is nevertheless reproduced in this chapter (Fig. 609, p. 668).

In 14 of the 20 cases of uncomplicated aortic stenosis, including two with subvalvular stenosis established on angiocardiography and at operation, respectively, distinct dilatation of the ascending aorta was visible (Fig. 593, a and b). In nine of them there was, in addition, a more localized in shape of the aorta. Thus, the ascending aorta describes a wide curve forward and to the right. In eight cases, the aorta showed no deformity (Fig. 596). In one of them, subvalvular stenosis was found at autopsy (Fig 597). No hypoplasia of the aorta was observed.

The pulsations in the ascending aorta were large, whether or not dilatation was present. The largest *pulsations were* visible in the suprabulbar segment.

The aortic arch is characterized by Brown (107) as small and inconspicuous in aortic stenosis. In our series, it was dilated in scarcely half the cases and of the ordinary appearance in the remainder, with one

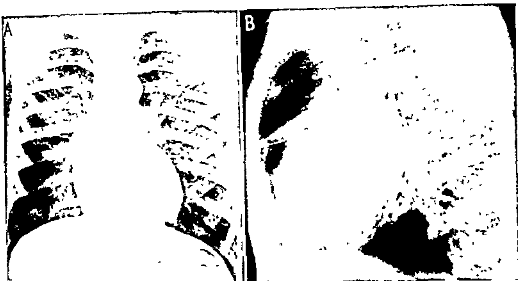


Fig 593a —Subvalvular aortic stenosis Boy, aged 10 (C W. 440427). Hypertrophy of left ventricle, with an increase in breadth of apex. Poststenotic dilatation of ascending aorta Enlargement of left atrium, caused by impediment to outflow to the hypertrophied left ventricle.

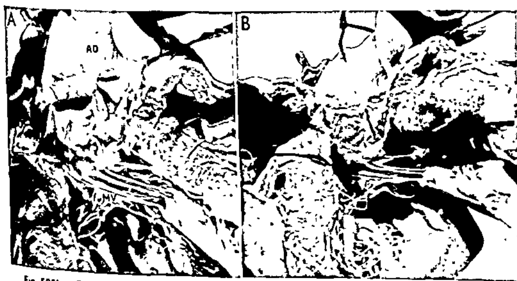


Fig 593b —Same case as in Figure 593a About 1 cm below the thickened but otherwise normal aortic cusps, circular constriction of the infundibulum by an irregular structure [illegible]
sembling a [illegible]
cusp and
below AC
nary cusp
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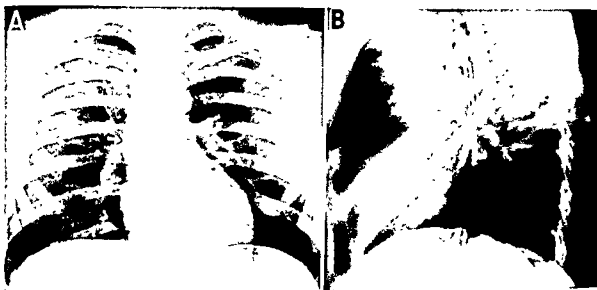


Fig. 594.—Aortic stenosis. Boy, aged 15 (B.W. 370329). Slight enlargement of left ventricle; appearance as in hypertrophy. Suprabulbar dilatation of ascending aorta, visible as a bulge in the outline (arrow in *A*). Left atrium normal, inappreciable increase in heart volume

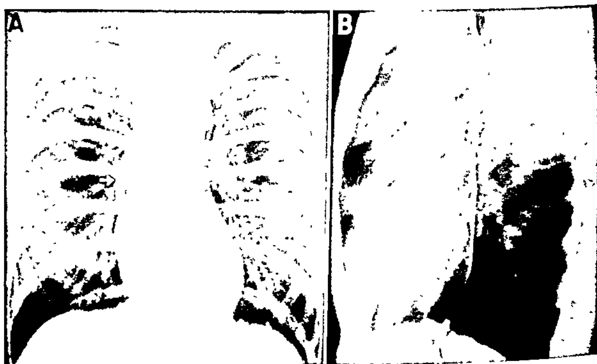


Fig. 595.—Valvular aortic stenosis Boy, aged 14 (V.S. 390318); see Figure 608. No enlargement of left ventricle, no increase in heart volume, elongation of ascending aorta, which is dilated, particularly in suprabulbar segment (arrow in *A*).



596.—Aortic stenosis Boy, aged 11 (C F 420207) Hypertrophy of left ventricle, no dila- of aorta, increase in heart volume.

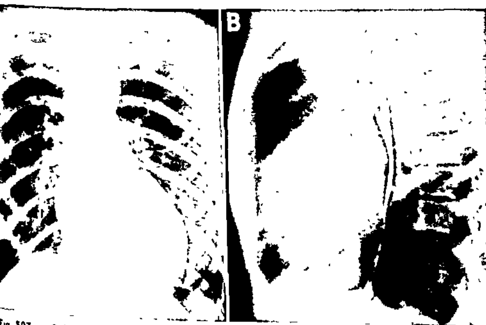


Fig 597a —Subvalvular aortic stenosis Girl, aged 14 (B D. 370625) Great hypertrophy and enlargement of left ventricle, distinct increase in heart volume, no or inappreciable dilatation of ascending aorta

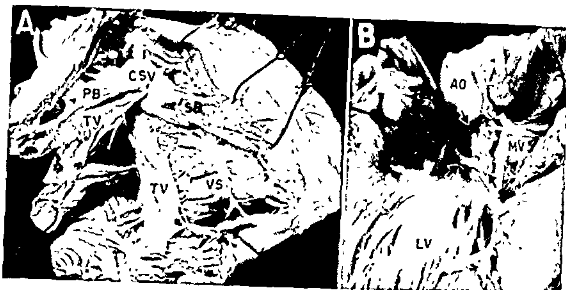


Fig. 597b.—Same case [illegible]
the membranous part of [illegible]
presses the lumen in a sem[illegible]
the septum and *anterior* cusp of the mitral valve and [illegible]
[illegible]
[illegible] the right ventricle (*A*), causing considerable curvature of
the septal band. *AO*, *aorta*, *CSV*, crista supraventricularis, *I*, infundibulum, *LV*, left ventricle, *MV*, mitral valve, *PB*, parietal band, *SB*, septal band, *TV*, tricuspid valve; *VS*, ventricular septum.

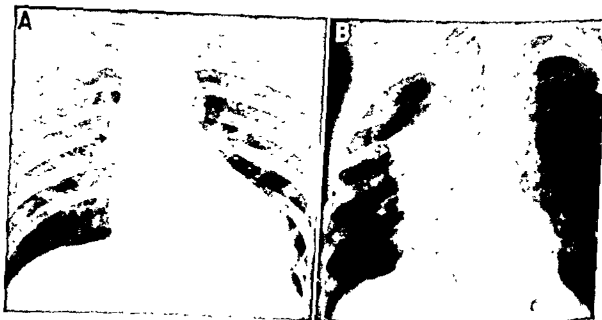


Fig. 598.—Aortic incompetence and stenosis Boy, aged 14 (L G 380119). Considerable hypertrophy and dilatation of left ventricle, gross dilatation of ascending aorta and aortic arch, *extending into superior* part of descending aorta. Dilatation is most conspicuous in the suprabulbar region (arrows). Distinct increase in heart volume

exception, in which it was narrow. Large pulsations were also observed over this segment of the aorta, and in 18 out of 20 cases an abnormal electrokymogram was recorded

The width and shape of the descending aorta were normal. The pulsations were of ordinary size, in contrast to the cases with coincident aortic incompetence, in which they were distinctly increased even in the dilated segment below the aortic arch.

No calcifications of the aortic orifice could be demonstrated, either on the roentgenogram or on fluoroscopy. In Campbell and Kauntze's series (137), the youngest patient with calcifications was 18 years old

The left ventricle was enlarged in barely half of our cases, the left and posterior part of its outline exhibiting increased curvature and elongation In some cases distinct curvature of the apex was visible. The enlargement was not marked and was presumably due mainly to hypertrophy. The heart volume was only moderately

the aortic isthmus.

The clinical observations gave reason for us to assume that stenosis was *more severe* in seven of the cases than in the remainder (see p 648). In one of the probably least advanced of these cases (Fig 595), *angiocardiography* showed the existence of valvular stenosis (Fig. 608), but the width of the orifice could not be determined No enlargement or change in shape of the left ventricle was found on either roentgenologic or angiocardiographic examination. The ascending aorta was greatly dilated, elongated, and curved The heart volume was *not* increased, this observation also applied in an additional two cases of severe stenosis

In a second case (Fig 596) in which angiocardiography with injection of contrast medium into the pulmonary artery was also performed, and in which the clinical findings indicated the presence of a severe degree of stenosis, it could not be determined whether it was valvular or subvalvular. The left ventricle was moderately enlarged and the heart volume increased to a corresponding degree, whereas the aorta had the usual width and appearance. Moderate enlargement of the heart was also present in one case of severe subvalvular aortic stenosis.

Considerable enlargement of the heart was seen in one case of subvalvular stenosis (Fig. 597), as well as in one case of severe stenosis complicated by coarctation of the aorta and anomalous pulmonary venous return.

In all of the other cases, in which stenosis was probably slight, the enlargement of the left ventricle and increase in heart volume were not conspicuous or were lacking In view of the fact that cardiac enlargement may be slight even in very severe stenosis and that sudden death may occur even when the heart is not enlarged—as in one of our cases—only a distinct increase in heart volume presumably is of any decisive prognostic importance in the individual case. No relation was found between the appearance of the aorta and the degree of stenosis

The cardiac configuration may not be typical if the aorta is not dilated and the change in shape of the left ventricle is not prominent In three of our cases the appearance was normal in every respect.

It is not possible to distinguish between valvular and subvalvular stenosis on the basis of the roentgenogram. Poststenotic dilatation of the aorta occurs in both conditions (96, 309, 479, 486), but it may be lacking in both Calcifications of the aortic orifice, which cannot as a rule be identified in children, argue in favor of valvular stenosis, but they have also been described in the subvalvular form. Nor does a study of the type of the aortic pulsations by means of electrokymography provide any decisive information

The roentgenologic features differed somewhat in three cases in which stenosis was combined with aortic incompetence (Figs 598 and 599). Thus, the dilatation of the aorta involved the superior segment of

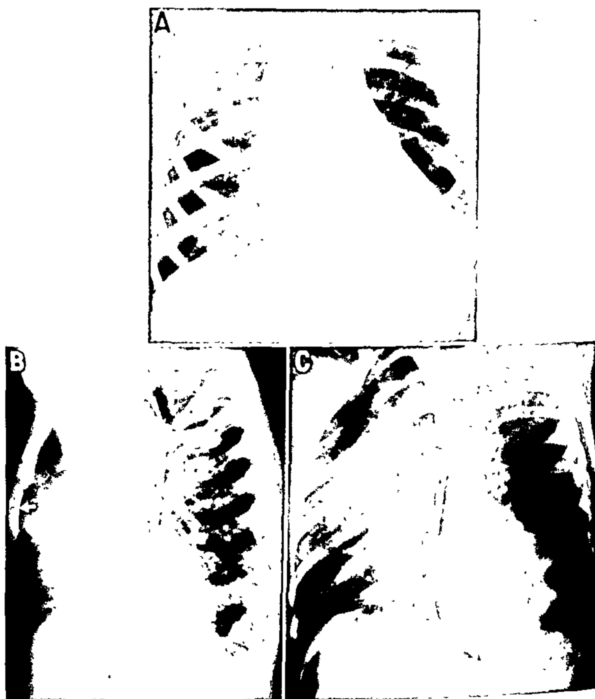


Fig. 599.—Aortic incompetence and stenosis. Boy, aged 6 (S T 470619), see Figure 613 Considerable increase in heart volume with great dilatation of left ventricle, marked widening of ascending aorta, particularly the anterior segment Arrow in *B* indicates a shallow indentation on dorsal surface of the sternum

the descending aorta as well, and the pulsations in it were also large. The ascending aorta was considerably dilated There were great dilatation of the left ventricle and lateral displacement of the apex in two of the cases In the third, the left ventricle was only moderately enlarged. The heart volume was increased to a corresponding degree

Aortic stenosis present as an associated malformation may be difficult to detect roentgenologically, particularly when in combination with another aortic anomaly None of the roentgenologic findings gave us reason to suspect its existence in two cases of patent ductus arteriosus in our series Nor could it be established in two cases of atrial septal defect with a left to right shunt at the atrial level

In certain respects, the roentgenologic findings pertaining to the respective sides of the heart are similar in aortic and in pulmonary stenosis Increased poststenotic pulsations are typical of both conditions Poststenotic dilatation nevertheless seems to be a more *constant* feature of pulmonary stenosis than of aortic stenosis, at any rate if the comparison is confined to the pulmonary valvular *form* Hypertrophy and possible enlargement of the relevant ventricle, with or without *an effect* on the total heart volume, is a characteristic feature of both aortic and pulmonary stenosis On the other hand, dilatation and hypertrophy of the right atrium are often found in moderately severe and severe pulmonary *stenosis*, whereas corresponding changes in the left atrium are seldom part of the picture in aortic stenosis

ELECTROKYMOGRAPHY

The occurrence of abnormal pulsations in the thoracic aorta could be established in all 16 cases studied

In *aortic stenosis*, the electrokymogram of the ascending aorta showed the following alterations The upper part of the systolic upstroke, which normally ends in a peak, was more or less rounded (Figs 600–605) or this limb rose more or less continuously (Fig 604) The incisura and protodiastolic phase were indistinct. The dicrotic wave was reduced variably and in some cases was merely suggested. Small vibrations were sometimes superimposed on the systolic limb (Figs. 603 and 605). An anacrotic notch was only exceptionally present and was never as distinct as in pulmonary stenosis. The appearance of the curves varied to some extent according to the segment of the vessel over which they were recorded (Fig 605). This variation was mainly of the same regular nature as that in tracings recorded in normal cases (p. 113). The alterations in the electrokymograms seemed to be essentially an expression of the prolonged emptying of the left ventricle and of the abnormal function of the aortic orifice.

In the curves from the aortic arch as well, changes were found in the configuration of the upstroke (Figs. 602–605). As a rule, the rise continued until the end of systole. A halt was sometimes present, and marked the transition into a somewhat slower upswing in the upper part. The incisura and dicrotic wave were usually diminished and were sometimes obliterated. In some cases, the diastolic limb had a curved course, convex upward

In the curves of both the ascending aorta and the aortic arch, the onset of systolic upstroke occurred without noticeable delay. The systolic click recorded in six cases on the phonocardiogram registered simultaneously over the apex appeared concurrently with the beginning of ejection in the aorta, or at most 0 03 sec later, as disclosed by the onset of upstroke on the ascending aorta electrokymograms (553)· see Figures 604 and 605

In two of the mild cases, the tracing of the aortic arch exhibited no significant changes, whereas that of *the* ascending aorta was definitely pathologic. In one case, the reverse applied.

In *severe* or moderately severe stenosis (see p 659 and Figs. 602–605), the systolic upstroke in *the* aortic arch curves was distinctly prolonged as compared with that in the mild cases It was nevertheless remarkable that in two of *the cases* of severe sub-

valvular stenosis the electrokymograms showed only moderate deviations from the normal. A continuous rise in the systolic upstroke on the ascending aorta curves was only exceptionally seen. These features are illustrated in Figure 605, in which the in- descending limb in the tracings from th aortic arch in the two most severe case was noteworthy.

Electrokymography does not seem to pro vide any means of differentiating betwee valvular and subvalvular stenosis. In a

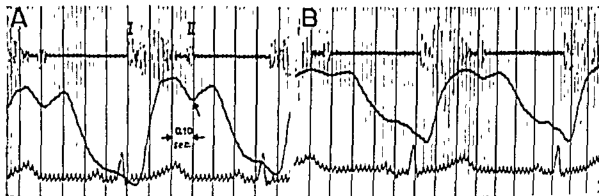


Fig. 600.—Electrokymograms in mild aortic stenosis. Boy, aged 10 (C.M. 440427). PCG ove aortic area. Time marking, 0 10 sec between thick lines. *A*, ascending aorta. Tracing practicall normal, but systolic upstroke ends without a distinct peak, gradient of upstroke normal. Incisur (arrow) corresponds to 2nd aortic sound *B*, aortic arch. Suggested rounding of upper part o contour of upstroke, protodiastolic phase preceding incisura somewhat ill-defined; no other de formities *I*, 1st sound, *II*, 2nd sound.

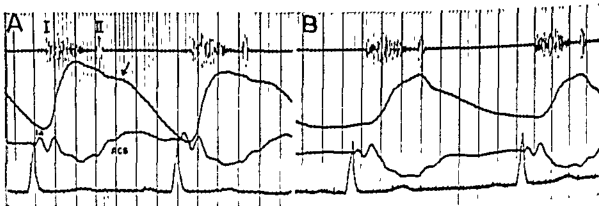


Fig. 601.—Electrokymograms in mild aortic stenosis. Girl, aged 16 (A.W. 380913). PCG ove [illegible] ascending aorta. Abnorma [illegible] wave low (arrow) [illegible] curve gives a false impression of decreased gradient), dicrotic wave obliterated. *I*, 1st sound, *II*, 2nd sound, *Ia*, pre sumed closure of mitral valve.

clination time is definitely prolonged, ranging from 0.16 to 0.20 sec in curves recorded from different segments of the aorta. Superimposed small vibrations in upstroke occurred only in the severe cases. The degree of reduction of the dicrotic wave could not be taken as a gauge of the degree of stenosis. The position of the wave far down on the

comparative series of 20 cases of acquired aortic stenosis, the electrokymograms showed the same changes in configuration as in the congenital cases.

It is known that in aortic *stenosis*, as in pulmonary stenosis, a suction effect occurs in the poststenotic segment at one stage in the phase of rapid ejection, although in-

verse movements of the vessel wall cannot be recorded either on border or on density electrokymograms. The probable reason is that the effect is not apparent in the peripheral segment of the vessel. The conditions at the actual site of the aortic root cannot,

reduction in the phase of isometric contraction. The same phenomenon was noted in curves recorded over the left ventricle (Fig [illegible]). The ascending aorta tracings were also characterized by a rapid fall from the summit. The incisura and greatly reduced

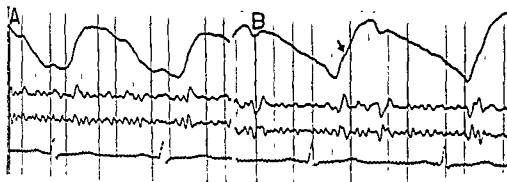


Fig 602 – Electro[illegible]
(V S 39[illegible]
Part of c[illegible]
Prominen[illegible]
[illegible] slight prolongation of systolic upstroke, distinct incisura but reduced dicrotic wave, diastolic limb slightly convex, anacrotic inflection (arrow).



Fig 603 —Electrokymograms in severe subvalvular aortic stenosis. Girl, aged 14 (B D 370625) *A*, ascending aorta Small vibrations superimposed [illegible] which has [illegible]
not delay[illegible]
discernib[illegible]
crotic wa[illegible]

however, be evaluated by electrokymography

In the three cases in which *aortic incompetence* predominated, the systolic upstroke showed the normal steep gradient in the ascending aorta curves As opposed *to* the conditions in isolated stenosis, onset of upstroke was early It was recorded at the latest 0 08 sec after the Q wave, indicating a

dicrotic wave occurred far down on the descending limb. Another finding was a presystolic upswing in the upstroke (Fig 607); it was probably due to the influence of auricular systole

In the electrokymograms from the aortic arch, the *incisura* and dicrotic wave [illegible]

valvular stenosis the electrokymograms showed only moderate deviations from the normal. A continuous rise in the systolic upstroke on the ascending aorta curves was only exceptionally seen. These features are illustrated in Figure 605, in which the in-

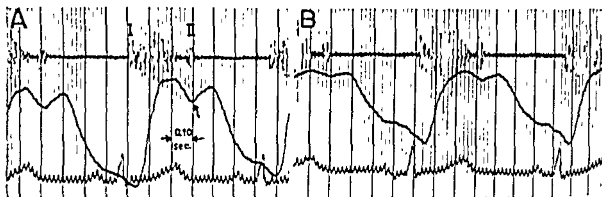


Fig. 600.—Electrokymograms in mild aortic stenosis. Boy, aged 10 (C M. 440427). PCG over ... ascending aorta Tracing practically ... Incisura ... part of ... other deformities. *I*, 1st sound, *II*, 2nd sound.

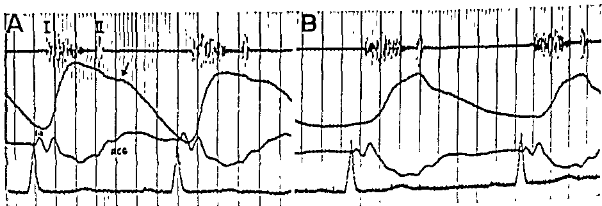


Fig. 601.—Electrokymograms in mild aortic stenosis. Girl, aged 16 (A.W. 380913). PCG over ... ascending aorta. Abnormal ... otic wave low (arrow). ... he curve gives a false impression of decreased gradient), dicrotic wave obliterated *I*, 1st sound, *II*, 2nd sound; *1a*, presumed closure of mitral valve

clination time is definitely prolonged, ranging from 0.16 to 0 20 sec in curves recorded from different segments of the aorta. Superimposed small vibrations in upstroke occurred only in the severe cases. The degree of reduction of the dicrotic wave could not be taken as a gauge of the degree of stenosis The position of the wave far down on the descending limb in the tracings from the aortic arch in the two most severe cases was noteworthy.

Electrokymography does not seem to provide any means of differentiating between valvular and subvalvular stenosis. In a comparative series of 20 cases of acquired aortic stenosis, the electrokymograms showed the same changes in configuration as in the congenital cases.

It is known that in aortic stenosis, as in pulmonary stenosis, a suction effect occurs in the poststenotic segment at one stage in the phase of rapid ejection, although in-

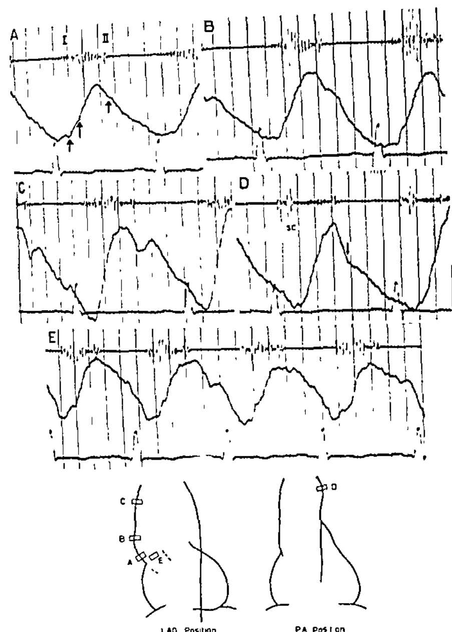


Fig 605 —Electrokymograms in severe aortic stenosis Boy, aged 11 (C F. 420207) PCG over

[illegible]

anacrotic notch, incisura distinct, dicrotic wave only moderately diminished. *D*, aortic arch PCG over the apex, shows systolic click (*sc*) Slow systolic upstroke, incisura lies far down, indistinct dicrotic wave *E*, density electrokymogram recorded above the aortic root. Distinct anacrotic notch, incisura and dicrotic wave cannot be identified The electrokymogram varies somewhat in the different cardiac cycles *I*, 1st sound, *II*, 2nd sound

protodiastolic phase preceding the incisura was poorly defined. The upstroke had the same appearance as in aortic stenosis. It is unquestionable that, in certain cases, the aortic arch tracings in both aortic stenosis and in aortic incompetence may be identical.

stenosis, although it may provide some information. In severe stenosis with marked left ventricular hypertrophy, the left atrial pressure is raised; this can be established by recording the pulmonary capillary venous pressure. Any marked rise in pressure

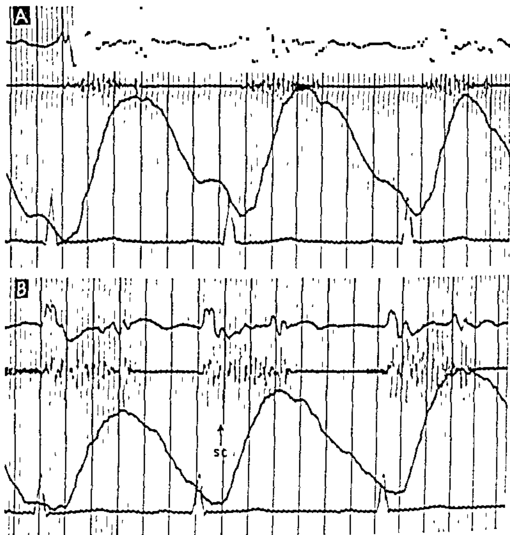


Fig. 604.—Electrokymograms in severe aortic stenosis and coarctation of the aorta. Boy, aged 9 (L.J 460810). PCG over aortic area in *A* and over apex in *B* Electrokymograms recorded over ascending aorta Onset of upstroke 0.13 sec after Q wave More or less continuous systolic rise. Incisura and dicrotic wave almost obliterated. Systolic click (*SC*) at onset of aortic systolic ejection

In two cases of severe aortic stenosis, the electrokymogram recorded over the left atrium was abnormal, indicating the presence of an impediment to atrial emptying

CARDIAC CATHETERIZATION

Catheterization of the right side of the heart is of little diagnostic value in aortic

in the pulmonary circulation is present only when there is left ventricular failure.

In order to establish the degree of aortic stenosis more exactly, it is necessary to determine the pressure gradient across the orifice, as well as the stroke volume of the left ventricle. If the foramen ovale is patent, it is possible to make direct measurements of the pressure in the left ventricle



Fig 608.—Valvular aortic stenosis Boy, aged 14 (V S. 390318). Injection into pulmonary artery Aortic cusps are f[illegible]
During diastole [illegible]
whole ascending [illegible]
and this segme[illegible]
Course of the aorta is [illegible]
fice not outlined AO

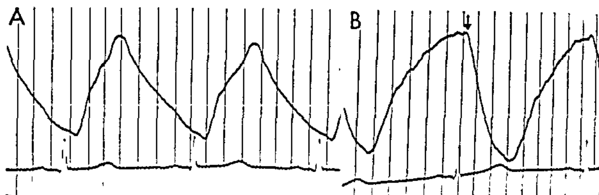
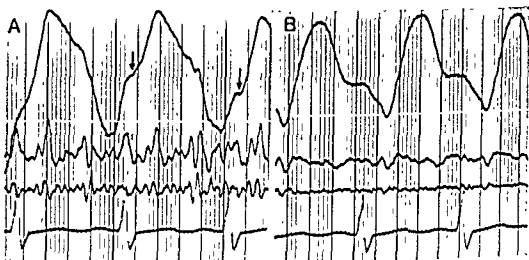


Fig. 606.—Electrokymograms in combined aortic incompetence and aortic stenosis. Boy aged 9 (K.H. 431007). *A*, aortic arch. Anacrotic notch, slow inclination in later part of upstroke probably representing prolongation of phase of reduced ejection, incisura and dicrotic wave obliterated. Tracing shows great similarities to that in Figure 603, *B*. *B*. left ventricle. Early systolic deflection, 0.06 sec after Q wave (arrow) indicating shortening of isometric contraction phase



[illegible] and aortic stenosis Boy, aged [illegible] orta. Presystolic upswing [illegible] 7 sec after the Q wave, it is steep and has inclination time of 0 08 sec. Rapid fall in second half of systole. Indistinct incisura and dicrotic wave *B*, aortic arch. Incisura and dicrotic wave lie far down on diastolic limb



g 608 —Valvular aortic stenosis Boy aged 14 (V S [illegible]) [illegible]
f Aortic cusps are fused into a do[illegible]
[illegible]

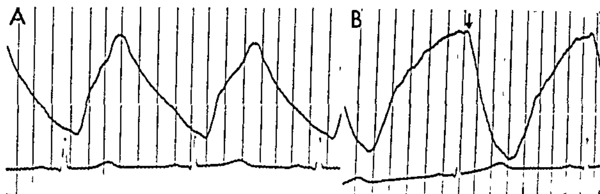


Fig. 606.—Electrokymograms in combined aortic incompetence and aortic stenosis. Boy aged 9 (K.H. 431007). *A*, aortic arch Anacrotic notch; slow inclination in later part of upstroke probably representing prolongation of phase of reduced ejection; incisura and dicrotic wave obliterated. Tracing shows great similarities to that in Figure 603, *B*. *B*, left ventricle Early systolic deflection, 0 06 sec after Q wave (arrow) indicating shortening of isometric contraction phase

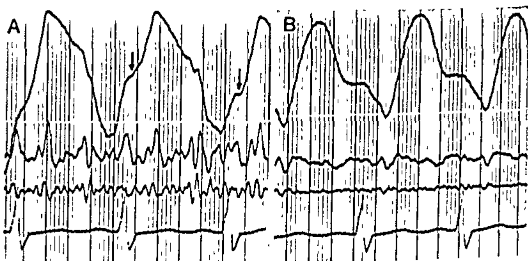


Fig. 607.—Electrokymograms in combined aortic incompetence and aortic stenosis Boy, aged 6 (S.T 470619). Frequency channels, 10 and 20 cps *A*, ascending aorta. Presystolic upswing probably due to atrial influence. The true upstroke (arrows) starts 0.07 sec after the Q wave, it is steep and has inclination time of 0.08 sec. Rapid fall in second half of systole Indistinct incisura and dicrotic wave *B*, aortic arch Incisura and dicrotic wave lie far down on diastolic limb



Fig. 609 (*cont*)

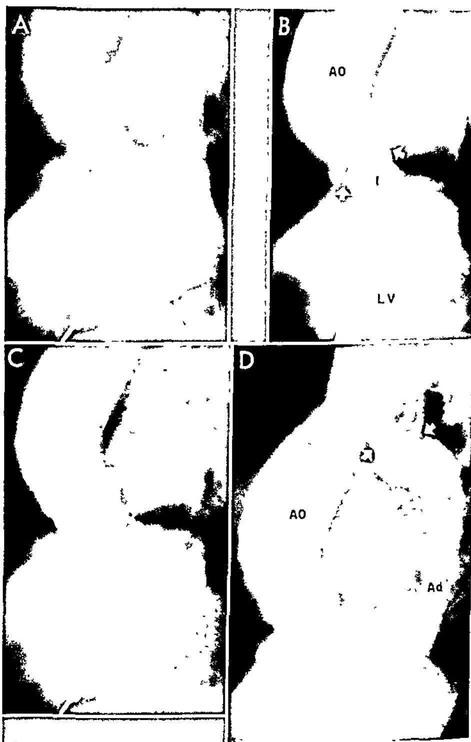


Fig. 609.—Valvular aortic stenosis and coarctation of the aorta *Girl, aged 11* (L C 460210). Perci [illegible] medium into this chamber. [illegible] lar p[illegible] Coarctation of the ao (*Ad*). Moderate post- (*continued*).

through a catheter introduced from the right side of the heart. As a rule, left heart catheterization must be performed. After puncture of the left atrium from the back (67) or transbronchially (9, 247), a narrow catheter is introduced through the needle and advanced into the left ventricle and thence into the aorta (32, 63). Another

formed in 11 of our cases. The pulmonary artery and pulmonary capillary venous pressure were found to be normal in all but one patient. This was a 16-year-old girl (E.I. 410918) with severe valvular stenosis, mild coarctation of the aorta, and nomalous pulmonary venous drainage, he veins from the right upper and middle bes opening into the superior vena cava see Angiocardiography). She died in connection with operation, and the diagnosis ould be verified at autopsy. On percuneous puncture of the left ventricle, a ystolic pressure of 279 mm Hg and an nd-diastolic pressure of 34 mm were recorded The pressure gradient across the rtic valve was 146 mm Hg The left to ght shunt was 2 8 liters per minute, with systemic flow of 4 0 (= 2 8 ml per m² S A.) and a pulmonary flow of 6 8 liters er minute. The left ventricular stroke volme was only 52 ml (= 36 ml per m² S A.) The mean pressure in the pulmoary artery was slightly raised (24 mm g), but the PCV pressure was not recorded In view of the high diastolic presre in the left ventricle, the left atrial ressure must also have been high Since e pressure in the right atrium was noral, the fall in pressure over the pulmoary vascular bed draining into the supeor vena cava was greater than that over e normally drained part. Bronchospiromry showed that the right lung was responble for a greater part of the total oxygen ptake than normally, indicating an increased blood flow through the right lung Percutaneous left ventricular puncture as performed in another two cases One patient was a 13-year-old boy with subvalvular stenosis, the left ventricular pressure was 185 mm Hg systolic and 15 mm end-diastolic. The other was a 5-year-old boy with valvular stenosis; the left ventricular pressure was 190/27 mm Hg.

ANGIOCARDIOGRAPHY

The essential prerequisite for angiocardiographic diagnosis of the nature of the impediment to outflow in valvular and subvalvular aortic stenosis, respectively, is satisfactory opacification of the left ventricle and aorta. In venous angiocardiography, the limitation of the method in this respect is particularly apparent. However, in our experience there are difficulties in demonstrating the anatomic changes even by means of the semiselective technique with injection of contrast medium into the pulmonary artery This is partly because in the frontal or oblique projections the opacified left atrium and pulmonary veins are completely or partly superimposed on the aortic root and outflow tract of the left ventricle. Only in the lateral view is the aortic orifice depicted without superimposition of these structures. We applied the semiselective technique in three cases of aortic stenosis, but the impediment could be localized on the basis of the angiocardiograms in only one of them (Fig 608). The width of the orifice could be determined neither in the frontal nor in the lateral views nor on complementary examination

dium directly into the left ventricle after puncturing it percutaneously. This selective method was later used by Lehman *et al* (421) in a large series, including cases of acquired aortic stenosis

We have made such examinations in aortic and mitral disease in collaboration with Drs. V. O Bjork, G. Malmström, and B Nordenström (65). Three of our cases of *congenital* aortic stenosis are included in this series It can be inferred from Figures



Fig. 610.—Subvalvular aortic stenosis Boy, aged 13 (K S 440512). Percutaneous puncture of left ventricle (*LV*) and direct injection of contrast medium In the infundibulum (arrows in *D*), about 1 cm below the
in *C*) is visible in systole, .
fundibulum and bulb of
aortic cusps are distinct,
the outline. (Courtesy of Dr. B. Nordenstrom, Thoracic Clinic, Karolinska Sjukhuset)

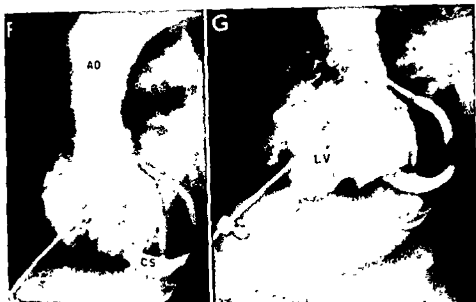


Fig 611 (*cont.*)



Fig 612 —Valvular aortic stenosis The cus... thick and partly calcified. *A*, view from left ver... wall muscles and trabeculae of left ventricle ... tricular septum

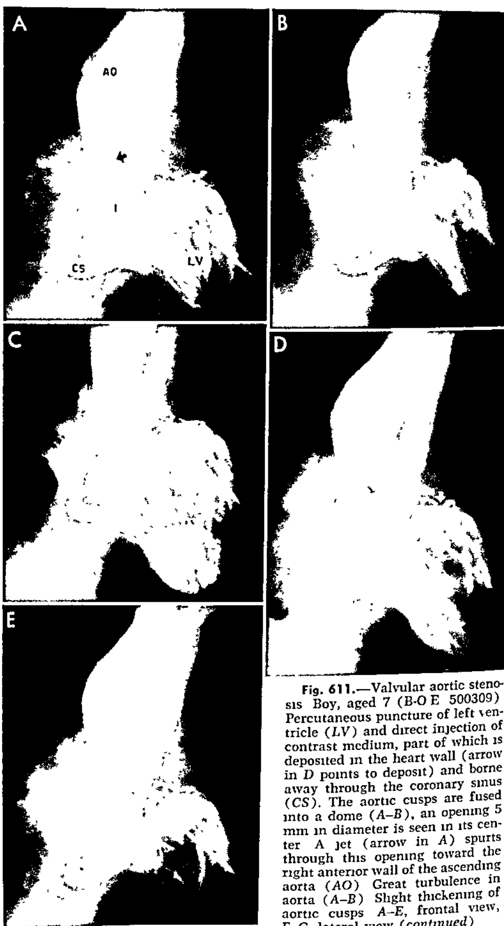


Fig. 611.—Valvular aortic stenosis Boy, aged 7 (B-O E 500309) Percutaneous puncture of left ventricle (*LV*) and direct injection of contrast medium, part of which is deposited in the heart wall (arrow in *D* points to deposit) and borne away through the coronary sinus (*CS*). The aortic cusps are fused into a dome (*A–B*), an opening 5 mm in diameter is seen in its center A jet (arrow in *A*) spurts through this opening toward the right anterior wall of the ascending aorta (*AO*) Great turbulence in aorta (*A–B*) Slight thickening of aortic cusps *A–E*, frontal view, *F–G*, lateral view (*continued*)

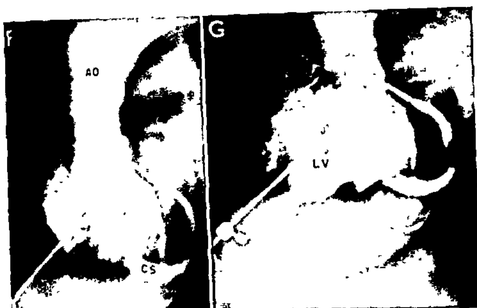


Fig. 611 (*cont.*)



Fig 612 —Valvular aortic stenosis The cusps, fused into a dome with a central orifice, are thick and partly calcified *A*, view from left ventricle *B*, view from aorta Great hypertrophy of wall muscles and trabeculae of left ventricle *MV*, mitral valve, *PM*, papillary muscle, *VS*, ventricular septum

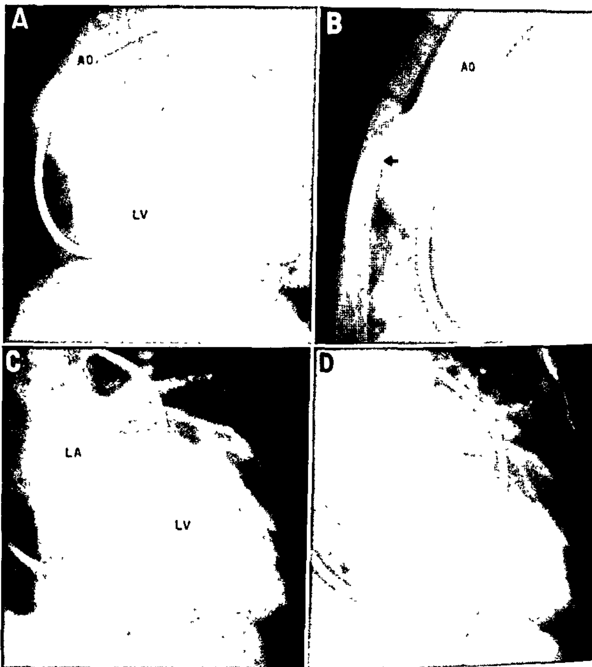


Fig. 613.—Aortic incompetence and stenosis. Boy, aged 6 (S T. 470619) Both left ventric and ascending aorta are greatly dilated At arrow in *B*, ascending aorta is locally dilated ar contiguous to the sternum. *AO*, aorta, *LA*, left atrium, *LV*, left ventricle

609, 610, and 611 that this method allows selective depiction of the left ventricle and aorta, so that the exact nature of the valvular or subvalvular stenosis can be established There are reasons to believe that even mild forms of the anomaly can be detected with this form of angiocardiography. In coarctation of the aorta with suspicions of associated valvular or subvalvular stenosis, this method seems to be particularly suitable for depicting both malformations on the same examination (Fig. 609).

In one of our cases with dominating aortic incompetence (Fig 613) in which the injection was made into the pulmonary artery, no stenosis of the aortic orifice could be visualized It could, however, be concluded that aortic incompetence was pres- [illegible] tween the ventricle and the aorta Both the ascending aorta and the left ventricle were greatly dilated.

21 Pulmonary Incompetence

WHEN THERE is a large flow through the pulmonary artery due to a left to right shunt, the orifice is often so greatly dilated that functional incompetence of the cusps arises. As an isolated malformation, pulmonary incompetence is extremely rare, but several cases have been reported in recent years (18, 493, 502) Congenital absence of the pulmonary valve has also been described (141).

Since the diastolic pressure in the pulmonary artery is normally low, incompetence of the cusps is not associated with such marked regurgitation as in corresponding damage to the aortic valve. Experimental studies have shown that dogs are well able to tolerate total extraction of the pulmonary valve during an observation period of 14 months (234).

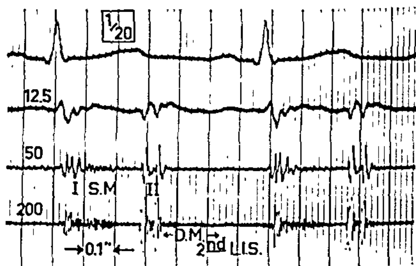


Fig. 614.—Phonocardiogram in pulmonary incompetence Boy, aged 7 (S A 441115) There is a faint systolic murmur (*SM*), duplicated 2nd sound (*II*) and faint protodiastolic murmur (*DM*) Boxed figures denote degree of amplification, which was high in this case, owing to faintness of the murmurs, other figures denote standard frequencies of the filters

Our series contains two patients with pulmonary valvular incompetence: a 7-year-old boy and a 9-year-old girl. They had never exhibited cardiac symptoms On physical examination, a long diastolic

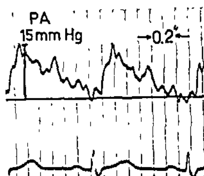


Fig 615 —Same case as in Figure 614 Pressure recording from the pulmonary artery shows low diastolic pressure

murmur was found, as well as a shorter systolic murmur with a maximum over the second left interspace. The second sound over the pulmonary area was duplicated (Fig 614). The electrocardiographic record was completely normal.

On *roentgenologic examination* no abnormalities were visible in the shape of the heart, and the appearance of the pulmonary artery was normal There was no pathologic increase in the heart volume

No shunts could be demonstrated on *cardiac catheterization* The pressure in the right ventricle was normal (20 2 and 32/8 mm Hg, respectively). The pulmonary artery systolic pressure was also normal (15 0 and 22/4 mm Hg, respectively). The diastolic level was abnormally low (Fig 615) A distinct notch was seen, followed by a fall in pressure to zero The leak through the pulmonary valve seemed to be insignificant and had not given rise to any enlargement or hypertrophy of the right ventricle The heart disease was diagnosed so late that it could not be stated with certainty to be congenital.

22 Ebstein's Malformation of the Tricuspid Valve

EBSTEIN'S DISEASE is characterized by deformity of the tricuspid valve. The anterior cusp is usually normally attached to the fibrous ring surrounding the atrioventricular orifice, but the other cusps are attached to the ventricular wall. The cusps are greatly deformed and displaced downward into the ventricle. The atrium is grossly dilated and forms an enormous chamber together with the supravalvular (proximal) region of the right ventricle.

Baker *et al.* (35) have stated—on the basis of studies on the normal development of the atrioventricular valves made by Odgers (512), among others—that the anomaly presumably arises during early embryonic development. It is due to inhibition of the normal regression and retraction of connective tissue and musculature in the valvular primordium. The displacement in the attachment of the cusps of the tricuspid valve can therefore be regarded as a direct result of this abnormal process. Some postmortem findings are illustrated in one of our cases.

[illegible]

and atrium The right ventricle rested like a cap on the dorsally situated, small left ventricle. The distance from the attachment of the cusps of the tricuspid valve to the apex was 6 cm, and from the diaphragmatic surface to the pulmonary valve, 10 cm Corresponding measurements in the left ventricle were 6 and 4 cm The part of the right ventricle below the valvular orifice (the distal part) was about five [illegible]

[illegible] cuspid valve were attached to the ventricular wall, 1 to 1.5 cm below the annulus fibrosus The anterior cusp was smaller than the others. The cusps were fused together and formed a network with clefts of varying size. This feature, together with the presence of numerous short chordae tendineae, which limited the mobility of the cusps, must have resulted in considerable incompetence The papillary muscles were small and at abnormal sites There was considerable hypertrophy of the trabeculae and their arrangement was abnormal. The actual ventricular wall was fairly thin (about 2 mm), this also applied to the region forming part of the right atrium. The trabeculae of the right atrium were greatly hypertrophied. Apart from a few minute defects (Fig. 629b, D), the atrial septum was intact The foramen ovale was closed. The left atrium was small The mitral orifice was normal and 1 5 cm in width

The size of the proximal and distal parts of the right ventricle varies appreciably. In most reports, the proximal part has been stated to be larger than the distal, and the former is almost invariably denoted as dilated and thin-walled (45, 238, 305, 719). Cases have, however, been described in which the distal part was also greatly dilated (68, 719), this observation agrees with the findings in our autopsy case.

Judging by the findings at autopsy, there is often incompetence of the tricuspid

he (719), but stenosis may also be pres- t (385, 425) Clinical signs of regurgita- n in the form of a pathologic venous lse and liver pulsations are lacking; the competence has therefore been regarded of minor importance and as producing signs until the development of heart ilure (68, 238). The distensible right rium, which is greatly dilated in Eb- ein's disease, allows regurgitation of a onsiderable volume of blood without the appearance of any symptoms from the systemic veins (719) In several cases, the shape of the pressure curve from the right atrium has been similar to that in tricuspid incompetence (94, 294, 600) A rise in pressure in the atrium without regurgitation to the systemic veins might occur during ventricular systole by means of contraction of the proximal part of the ventricle The wall in this part of the ventricle is nevertheless thin and is presumably of subordinate importance in this respect

The explanation of the dilatation of the distal part of the ventricle, such as was seen in our autopsy case, appears to be tricuspid incompetence

The malformation results in the right ventricle having difficulty in maintaining an adequate pulmonary flow. The pressure in the right atrium is raised and, if the foramen ovale is patent, a right to left shunt and cyanosis occur.

Some patients have lived to a considerable age (53, 169), but many have died in childhood. Sudden death has also been reported

TABLE 19 —Ebstein's Disease. Symptoms and Signs in 6 Cases

Case	Cyanosis Onset	Symptoms	Physical Signs
.S. 470825 (girl, 7 yr.)	On exertion, at 1 yr	From 5 yr, easily tired, can walk fairly far, dyspnea after running	No precordial bulge, parasternal lift, faint systolic murmur over apex, distinct gallop rhythm over apex, double 2nd sound over 2nd L.I S
J 450106 (girl, 7½ yr)†	Slight on exertion, at 6 mo	Moderately disabled, never able to play like a healthy child	Slight precordial bulge, systolic thrill over apex, *moderately* loud systolic murmur over apex; sounds of normal intensity, opening snap of tricuspid valve, liver enlargement
N 451026 (girl, 8 yr)	Slight, at birth	Rapidly dyspneic and tired on exertion, no more than 3 flights of stairs	No precordial bulge, faint systolic murmur + short protodiastolic murmur over apex; normal intensity of sounds, opening snap of tricuspid valve
S.N 410721 (girl, 12 yr)‡	Moderate, at 7 yr	Increasingly disabled since age 7, increased fatigability, dyspnea at rest, walks maximum of 50 m	Precordial bulge, parasternal lift, systolic *thrill and very loud, harsh systolic murmur*, maximum over apex; faint diastolic murmur over apex, presystolic murmur, acc. 1st sound, 2nd sound inaudible
L-J.L. 411229 (boy, 15 yr)	0	None	No precordial bulge, parasternal lift, acc. 1st sound, pansystolic murmur over 4th L.I S
B.C. 440203 (boy, 12 yr)	Moderate, at birth	Since birth attacks of paroxysmal tachycardia, moderately disabled, dyspneic and tired on slight exertion	[illegible]

†Died one year later of heart failure ‡Died 16 months later of heart failure

It was formerly considered difficult, if not impossible, to diagnose this disease *in vivo* During recent years, however, several reports have appeared with detailed descriptions of a characteristic [illegible] (68, [illegible] 393, [illegible] 719).

Our series contains six cases of Eb

22 Ebstein's Malformation of the Tricuspid Valve

EBSTEIN'S DISEASE is characterized by deformity of the tricuspid valve. The anterior cusp is usually normally attached to the fibrous ring surrounding the atrioventricular orifice, but the other cusps are attached to the ventricular wall. The cusps are greatly deformed and displaced downward into the ventricle. The atrium is grossly dilated and forms an enormous chamber together with the supravalvular (proximal) region of the right ventricle.

Baker *et al.* (35) have stated—on the basis of studies on the normal development of the atrioventricular valves made by Odgers (512), among others—that the anomaly presumably arises during early embryonic development. It is due to inhibition of the normal regression and retraction of connective tissue and musculature in the valvular primordium. The displacement in the attachment of the cusps of the tricuspid valve can therefore be regarded as a direct result of this abnormal process. Some postmortem findings are illustrated in one of our cases.

GIRL, AGED 9 YEARS (I J. 450106, Fig 629b)—Autopsy showed a greatly enlarged heart, with enormous dilatation of the right ventricle and atrium The right ventricle rested like a cap on the dorsally situated, small left ventricle. The distance from the attachment of the cusps of the tricuspid valve to the apex was 6 cm, and from the diaphragmatic surface to the pulmonary valve, 10 cm Corresponding measurements in the left ventricle were 6 and 4 cm The part of the right ventricle below the vular orifice (the distal part) was about [illegible]

cuspid valve were attached to the ventri[illegible] wall, 1 to 1.5 cm below the annulus fibr[illegible] The anterior cusp was smaller than the ot[illegible] The cusps were fused together and form[illegible] network with clefts of varying size This [illegible]ture, together with the presence of nume[illegible] short chordae tendineae, which limited mobility of the cusps, must have resulte[illegible] considerable incompetence. The papillary [illegible]cles were small and at abnormal sites T[illegible] was considerable hypertrophy of the trabec[illegible] and their arrangement was abnormal. Th[illegible]tual ventricular wall was fairly thin (abo[illegible] mm), this also applied to the region form[illegible] part of the right atrium. The trabeculae of right atrium were greatly hypertrophied. A[illegible] from a few minute defects (Fig 629b, *D*), atrial septum was intact. The foramen o[illegible] was closed The left atrium was small. mitral orifice was normal and 1 5 cm in wi[illegible]

The size of the proximal and distal p[illegible] of the right ventricle varies appreciably most reports, the proximal part has b[illegible] stated to be larger than the distal, and former is almost invariably denoted as [illegible]lated and thin-walled (45, 238, 305, 71[illegible] Cases have, however, been described which the distal part was also greatly [illegible]lated (68, 719), this observation agr[illegible] with the findings in our autopsy case.

Judging by the findings at autopsy, th[illegible] is often incompetence of the tricus[illegible]

be misinterpreted as mitral valve disease. The similarity was particularly evident in three of our cases in which an extra sound reminiscent of an opening snap was heard over the apex (Fig. 616, *B* and *D*). Similar observations have been reported by others (72, 484) According to Grob *et al.* (305), the heart sounds are faint, this observation did not apply in our cases

The absence of a parasternal lift in combination with cyanosis has been regarded as characteristic of Ebstein's disease (484). Our case S N. 410721 nevertheless presented a marked parasternal lift and a precordial bulge. Right ventricular hypertrophy was evident on the ECG. Moreover, the systolic murmur was considerably louder than in the other cases (Fig 616, C) Since no other cardiac malformation could [illegible]

[illegible] in one case (Fig 616, *A*) a loud sound was audible in midsystole. Its occurrence, as well as that of the diastolic extra sounds, is presumably best explained on the grounds of the pathologic changes in the movements of the tricuspid valve cusps

Peripheral edema was present in none of our cases, but one patient had an enlarged liver She died one year after being examined by us, cardiac failure had developed rapidly, with massive edema and a pathologic venous pulse Another patient died of heart failure 16 months after the examination.

Electrocardiography

Cardiac arrhythmias are extremely common in Ebstein's disease The Wolff-Parkinson-White syndrome was present in one of our cases and has also been reported in the literature (108, 290, 294), and attacks of paroxysmal tachycardia are frequent occurrences In our series, no other form of congenital heart disease was associated with such a high incidence of arrhythmias in the course of cardiac catheterization The ECG is characteristic Figure 617 shows the electrocardiograms in two of our cases.

The greatly dilated, hypertrophied right atrium results in tall, broad P waves, particularly over the right precordium. The P-Q interval is usually prolonged. In the patient who died, autopsy disclosed a greatly dilated, thin-walled right ventricle. As in right ventricular dilatation caused by a large atrial septal defect, an M-shaped QRS complex is present, although it is still more marked in Ebstein's disease. Yater and Shapiro (719) found, in histologic examinations, that the right bundle branch was not interrupted. Both R and R_1 are low over the right precordium Since the right ventricle works under normal pressure, right ventricular hypertrophy should not occur. One of our patients (S.N. 410721) nevertheless exhibited both electrocardiographic and physical signs of right ventricular hypertrophy. Severe tricuspid incompetence is associated with an increased burden on the ventricle, and hypertrophy could arise on these grounds. In the case in question, the pressure curves from the right atrium and right ventricle appeared almost identical, only the amplitude being lower in the atrium Autopsy 16 months later disclosed severe tricuspid incompetence, as well as marked muscular hypertrophy of the small, distal part of the right ventricle

ROENTGENOLOGIC EXAMINATION

Even a few years ago, the roentgenologic appearance in Ebstein's disease was regarded as uncharacteristic and consequently as of little value for diagnostic purposes (107, 650). Later investigations have shown that the appearance is typical, so that difficulties are seldom encountered in making a diagnosis (294). When the roentgenogram is judged against the background of the clinical findings, the diagnosis can be considered as established in the majority of cases

The salient features are

Enlargement of the right atrium and right ventricle

Sparse vascularity of the lungs and, in most

stein's disease, all diagnosed *in vivo*. In addition, a roentgenologic examination was made of one adult patient from another hospital.

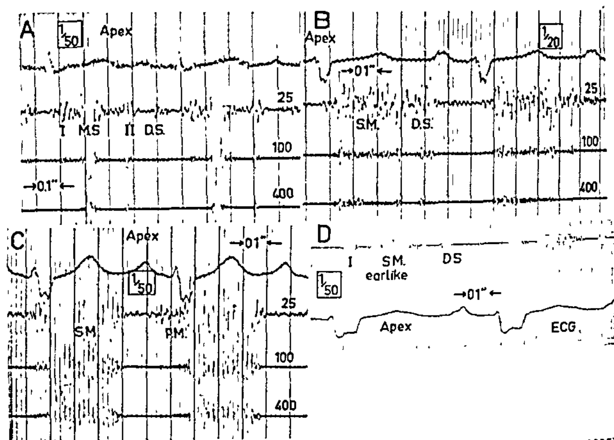
CLINICAL FEATURES

The chief symptoms and signs in our cases are assembled in Table 19. More than slight cyanosis was present in only two cases and had appeared fairly late in one of them

All but one of our patients were distinctly disabled. The most common symptoms were increased fatigability and dyspnea. One patient, a 15-year-old boy (L-J L. 411229), had no symptoms, but the exercise tolerance test showed moderately decreased working capacity (550 kgm per minute).

PHYSICAL SIGNS

Physical signs are by no means characteristic. A systolic murmur over the apex (Fig. 616) is the most constant finding and has been reported in practically all the published cases Tricuspid incompetence seems to be the most plausible explanation for the murmur Since the cusps of this valve lie far to the left, the sound phenomenon is heard and recorded best over the apex. A diastolic murmur appears to be more uncommon but was audible in most of our cases. In two it was presystolic (Fig. 616, C). Consequently, the condition is apt to



[illegible] cases of Ebstein's disease. *A*, girl, aged 7 (L S. 470825). [illegible] ly faint diastolic sound (*DS*) recorded over the apex. *B*, [illegible] nurmur (*SM*) and an extra diastolic sound (*DS*) recorded [illegible] 721) A very loud systolic murmur (*SM*) and faint pre- [illegible] girl, aged 8 (I J. 450106) A systolic murmur [illegible] the apex Boxed figures denote degree of ampli- [illegible] s of the filters.

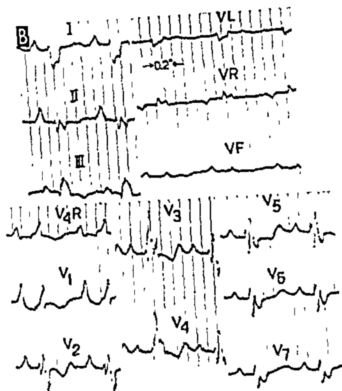


Fig 617 (*cont.*).—*B*, girl, aged 8 (I J 450106) High, peaked P waves over right precordium. Right bundle-branch block with probable right ventricular hypertrophy.



Fig 618.—Ebstein's disease Girl, aged 6 (L S 470825) Considerable dilatation of right atrium and ventricle Long contiguity of surface of ventricle to anterior wall of the thorax. In *A*, the main trunk of the pulmonary artery can be identified (arrow) Centrally and peripherally, great reduction in vascularity of the lungs Aorta is narrow

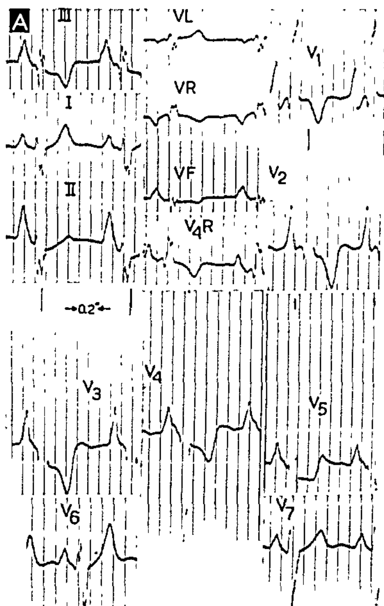


Fig. 617.—Electrocardiograms in Ebstein's disease. *A*, girl, aged 12 (S N. 410721). High, peaked P waves, predominantly over right precordium. Right ventricular hypertrophy and right bundle-branch block, with secondary S–T and T changes over right precordium (*continued*).

cases, lack of the normal prominence of the pulmonary artery

As a rule, a narrow aorta

We made roentgenologic examinations of seven patients · six children and one adult.

Enlargement of the right atrium was considerable and sometimes enormous (Figs. 618–622). All parts of the atrium, including the appendage, were dilated. Since the dilatation was in the forward, upward, and lateral directions, there was a great increase in the contiguity of the surface to the wall of the thorax. In some cases, the atrium extended to the level of the aortic arch, and its lateral border was long and curved. The dilatation also involved the posterior segment of the atrium, which usually formed the whole or the greater part of the posterior border of the heart in the lateral projection. Angiocardiography and cardiac catheterization showed that the valvular plane lay far to the left

The pulsations over the atrium are usually small, a feature that has been par-

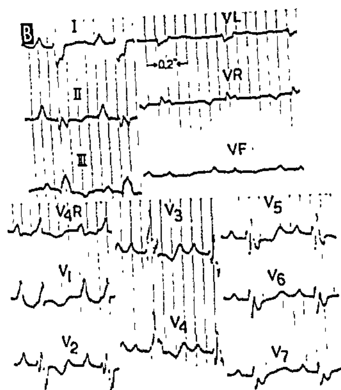


Fig 617 (*cont*) —*B*, girl, aged 8 (I J 450106) High, peaked P waves over right precordium ght bundle-branch block with probable right ventricular hypertrophy



Fig. 618 —Ebstein's disease Girl, aged 6 (L S 470825). Considerable dilatation of right atrium and ventricle Long contiguity of surface of ventricle to anterior wall of the thorax. In *A*, the main trunk of the pulmonary artery can be identified (arrow). Centrally and peripherally, great reduction in vascularity of the lungs Aorta is narrow.

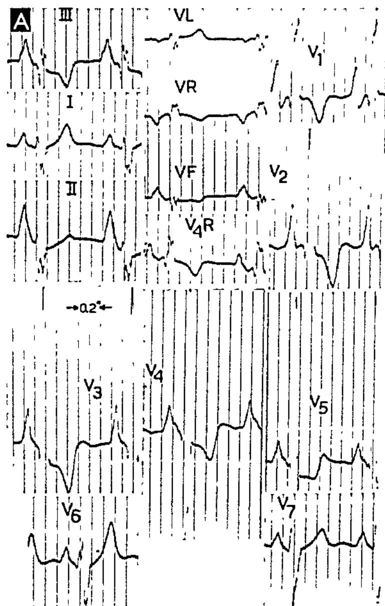


Fig. 617.—Electrocardiograms in Ebstein's disease. *A*, girl, aged 12 (S N. 410721) High peaked P waves, predominantly over right precordium Right ventricular hypertrophy and right bundle-branch block, with secondary S–T and T changes over right precordium (*continued*).

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The pulsations over the atrium are usu ally small, a feature that has been pa



Fig 621.—Ebstein's disease Girl, aged 12 (S N 410721). Right atrium and ventricle are still more dilated than in Figures 618–620, so that the shape of the heart differs to some extent

ticularly stressed by Engle *et al.* (238) Increased presystolic pulsations were, however, observed by Blacket *et al* (68) and by Wright *et al* (717) Among the factors on which the size of the pulsations depend are the volume of the atrium, the degree of tricuspid incompetence, the end-diastolic pressure in the right ventricle, and possible cardiac failure and atrial arrhythmia Consequently, individual variations are to be expected

In our patients, the pulsations were not enlarged, and in three cases they were distinctly small. The electrokymogram showed no increased presystolic activity. There was strikingly early systolic expansion of the atrial wall in four of them (see p. 687).

The infundibulum of the right ventricle was greatly dilated. In advanced cases the sinus region also was so markedly dilated that the whole of the left border of the heart

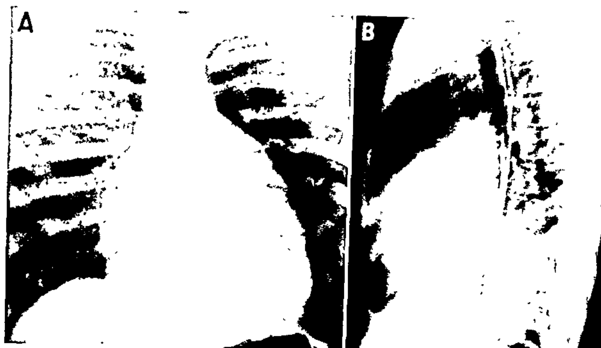


Fig. 619.—Ebstein's disease. Girl, aged 8 (I.N. 451026). Dilatation of right atrium and ven tricle more marked than in Figure 618. Infundibulum is so dilated that it overlaps the whole mai trunk of the pulmonary artery Greatly reduced vascularity of lungs, very narrow aorta.

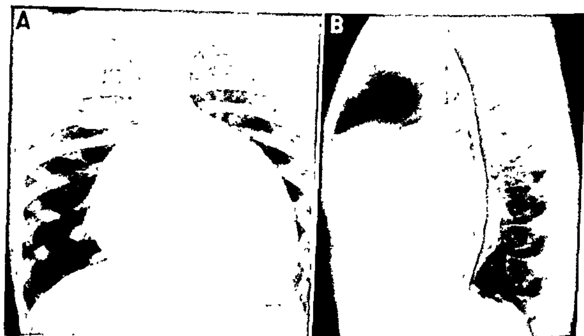


Fig. 620.—Ebstein's disease Girl, aged 9 (I J 450106). see Figure 629 Enormous dilatation of right atrium and ventricle, forming the whole anterior part of the heart In *B*, the atrial appendage fills the space between infundibulum and anterior thoracic wall Left ventricle and atrium, greatly decreased in volume, lie dorsally Marked reduction in vascularity of the lungs, aorta extremely narrow.



Fig 621 —Ebstein's disease Girl, aged 12 (S N. 410721) Right atrium and ventricle are still more dilated than in Figures 618–620, so that the shape of the heart differs to some extent

ticularly stressed by Engle *et al.* (238). Increased presystolic pulsations were, however, observed by Blacket *et al* (68) and by Wright *et al* (717). Among the factors on which the size of the pulsations depend are the volume of the atrium, the degree of [illegible] d[illegible] p[illegible] r[illegible] Consequently, individual variations are to be expected

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The infundibulum of the right ventricle was greatly dilated In advanced cases the sinus region also was so markedly dilated that the whole of the left border of the heart

was prolonged, curved and displaced laterally (Figs. 619–622). The anterior surface of the heart showed extensive contiguity to the thoracic wall, caused by both the right atrium and the right ventricle.

The pulmonary artery usually lacks prominence; comparisons with the angiocardiograms show that it is largely masked by the greatly dilated infundibulum. Pulmonary artery electrokymograms could, however, be recorded in all five patients examined, although in the advanced cases only tracings over a short segment were obtained. The caliber of the pulmonary artery cannot be evaluated by ordinary roentgenologic examination, and only in the mildest of our cases (Fig 618) did it appear distinctly. The pulsations were small. The vascularity of the lungs was extremely sparse, in several cases it was grossly reduced. Vascularity has been stated to be normal in less advanced and in mild cases (68, 294).

In most cases the aorta is narrow. As a rule, only the superior segment of the ascending aorta is visualized, owing to overlapping by the enlarged right atrial appendage. In extreme cases (Fig 622) the arch is the only part of the aorta to be projected above the enlarged heart. The impression in the esophagus made by the narrow aortic arch is shallow.

Enlargement of the heart is an almost constant feature, and the heart volume may amount to three to four times the normal volume.

In all seven of our cases, the roentgenogram was characteristic of Ebstein's disease. The individual differences in the outline are mainly to be ascribed to the variation in size of the right atrium and ventricle. The most advanced form of the malformation was seen in the adult patient and the least pronounced changes in the youngest patient

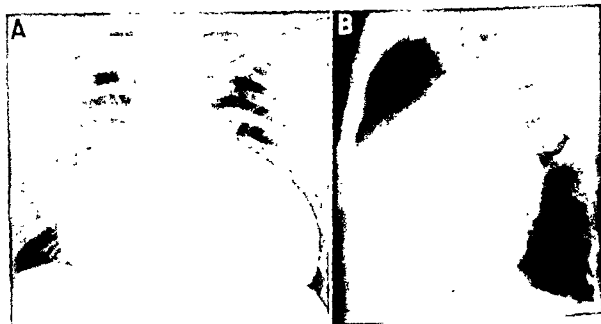


Fig. 622.—Ebstein's disease. Man, aged 42 (M.S. 120124). Shape of the heart resembles that in Figure 621. Dilatation of the right atrium and ventricle is still more pronounced (see Figs 618–621).

A similar roentgenologic appearance, characterized by dilatation of the right atrium and ventricle, decreased prominence of the pulmonary artery and reduced vascularity of the lungs, is seen in valvular pulmonary stenosis with heart failure. The poststenotically dilated pulmonary artery is then masked by the greatly dilated infundibulum.

From the purely roentgenologic standpoint, the appearance also shows great simi-

to that in exudative pericarditis. : differences are nevertheless pres- exudative pericarditis, the pulsa- e exceedingly small over all parts heart. It is true that in Ebstein's these pulsations may be small e right atrium, but they are of linary size over the left ventricle er, a narrow aorta is not a feature ertains to exudative pericarditis

emptying. These changes in the shape of the pulsations were interpreted as an effect of tricuspid incompetence. For some reason this did not appear in the tracings from the auricular appendage, which were of normal appearance

In two other cases (Fig. 625), a marked upstroke occurred before the Q wave in the right atrial tracing. The upstroke was followed, shortly after the R wave, by a deep

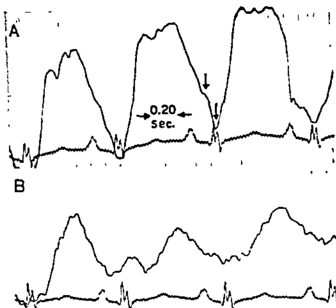


Fig 623 —Electrokymograms of right atrium in Ebstein's disease Girl, aged 8 (I N 451026). ody of atrium Curve is characterized by an early upstroke, starting at QRS, a plateau shape ing ventricular systole and an unimpeded phase of emptying No prolongation of atrial sys- (between arrows) B, auricular appendage Normal tracing

ELECTROKYMOGRAPHY

A study was made of five cases. Particu- interest was focused on the pulsations er the right atrium Their appearance ried somewhat in every case, and varia- ns also occurred in *different parts* of the rium. No increased presystolic activity as recorded

In two cases (Figs 623 and 624) the ain upstroke in the atrial tracing started t QRS in the ECG, and the course was ightly peaked or horizontal during ven- ricular systole. In early diastole, it was ucceeded by a rapid and deep phase of

deflection Similar curves were recorded over most segments of the atrium They could not be explained on the basis of tricuspid incompetence and must presumably be ascribed to abnormal movements of the atrium as a whole The possibility of such positional effects has been pointed out by Soloff *et al* (617)

In a third case, there was a complete difference between the shape of the electrokymogram of the auricular appendage and of that recorded over the segment which possibly corresponded to the part of the right ventricle incorporated with the atrium (Fig 626) The former showed *an ordinary*

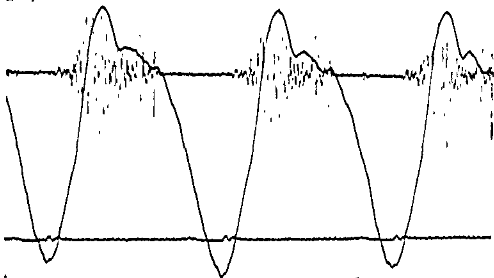


Fig. 624.—Electrokymogram of the right atrium in Ebstein's disease. Boy, aged 15 (L-J.L 411229). PCG recorded over tricuspid area. The curve is characterized by an early upstroke, starting at QRS, it ends in early systole in a peak, coinciding with the maximum of the systolic murmur. During the rest of systole, a suggested plateau, in diastole, a marked emptying phase

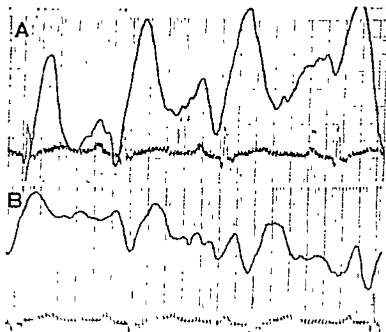


Fig. 625.—Electrokymograms of right atrium in Ebstein's disease Girl, aged 7 (L.S 470825). *A*, body of atrium Main upstroke starts before Q, early-systolic collapse, atrial systole of normal duration. *B*, auricular appendage Curve has the same general appearance as that in *A*.

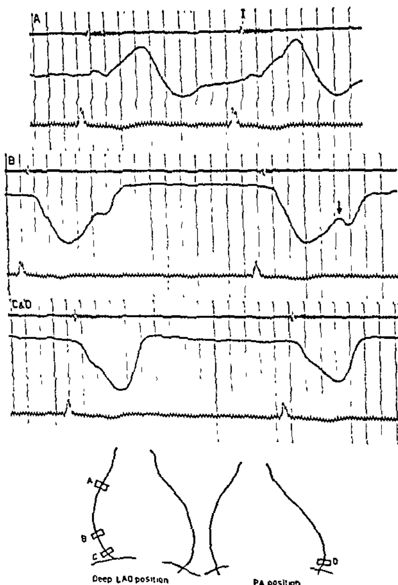


Fig 626 —Electro...
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filling phase during ventricular systole, whereas the latter had the shape of an abnormal ventricular curve and was characterized by a rapid filling phase in early diastole, ended abruptly by a limb with a lengthy horizontal course. The appearance was thus the same as that characterizing the ventricular electrokymogram in constrictive pericarditis. Similar curves were recorded over the apex, which belonged to the right ventricle. The curves indicate rapid dilatation of the ventricle, and of the part of it incorporated with the atrium, to maximal filling, and of its persistence for the rest of diastole. The pressure curves were also in agreement with those usually recorded in constrictive pericarditis (Fig 627).

The pulmonary artery electrokymograms were slightly abnormal in three cases, but the alterations were entirely uncharacteristic.

CARDIAC CATHETERIZATION

The performance of cardiac catheterization in Ebstein's disease is associated with severe cardiac arrhythmias, particularly when the catheter passes through the tricuspid orifice Continuous supervision of the electrocardiogram is therefore of prime importance Some authors have stated that the examination is associated with great risks for the patient (393, 459, 502)

On the basis of the position of the catheter, it is possible to demonstrate conclusively the enormous enlargement of the right atrium and the abnormal position of the tricuspid orifice The catheter coils up

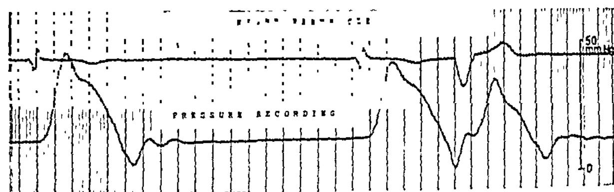


Fig. 627.—Man, aged 42 (M.S. 120124). Pressure curve from lower part of right atrium, proximal to cusps of the tricuspid valve. Appearance is the same as in constrictive pericarditis Systolic pressure 48 mm Hg, diastolic 15.

TABLE 20.—EBSTEIN'S DISEASE: RESULTS OF CARDIAC CATHETERIZATION IN 6 CASES*

Case	O₂ Content, vol. %							Arterial O_2 Sat. %	O_2 Cap., vol. %	Pressure, mm Hg				
	SVC	IVC	RA	RV	PA	PV	LA			RA Mean	RV Syst	RV End-diast	PA Syst	PA Diast
S N 410721	12.2	11.7	11.9	10.8	—	—	—	85	23.4	15	33	9	27	17
I.J 450106	11.3	11.1	10.9	10.2	10.4	—	—	90	22.2	14	36	6	24	7
I N 451026	10.3	11.6	11.3	11.3	11.3	—	—	94	19.1	13	23	8	24	8
L S 470825	10.0	—	10.5	10.6	10.6	—	—	81	19.5	—	14	5	13	7
L-J L 411229	11.4	12.8	11.4	11.6	11.3	—	—	97	17.6	4	16	4	13	4
B C 440203	12.9	16.0	14.4	14.8	14.6	20.1	17.9	90	20.5	6	33	7	28	9

*For abbreviations see Table 1, p 119

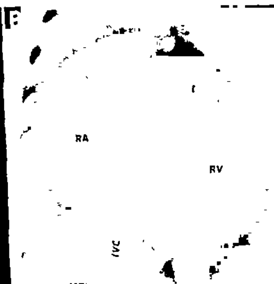
in the atrium and the borderline between it and the right ventricle can be recognized. In the frontal projection, this borderline appears abnormally far to the left. The right ventricle lies anteriorly, as usual, and the infundibulum is also at the ordinary site.

be done in our six cases. The results are shown in Table 20. In all but the mildest case, the pressure in the right atrium was increased, and in four cases *a rise in pressure* occurred during ventricular systole, indicating *tricuspid incompetence. The end-*



Fig 628 —Ebstein's disease. Girl, aged 8 (I.N. 451026). Pressure recordings from pulmonary artery (*PA*), right ventricle (*RV*) and right atrium (*RA*) Atrial and ventricular curves are strikingly similar, and no distinct transition can be seen on the withdrawal curve. Atrial curve shows signs of tricuspid incompetence





ig 629a.—Ebstein's disease Girl, aged 8 (I J 450106) Contrast medium was injected into t atrium Right atrium and ventricle are enormously dilated On the serial exposures, it can seen how the highly diluted contrast medium flows back and forth between the two chambers detaile ...

nay be difficult to obtain a pressure reding from the right ventricle. The catheis apt to be repelled into the atrium ing ventricular systole Arrhythmia may netimes prevent completion of the reding

Complete catheterization of the right side the heart and the pulmonary artery could

diastolic pressure was high in the right ventricle The systolic rise in pressure had a distinct appearance, with a short phase of ejection No sudden change in the shape of the curve was seen on withdrawal of the catheter from the ventricle to the atrium (Fig 628)

Decreased arterial oxygen saturation was

present in four patients, probably due to a small right to left shunt through a patent foramen ovale, verified in one case. One of these patients had become cyanotic at the age of 7 years. The reason seemed to be successive dilatation of the right atrium, the foramen ovale then became larger, with a concurrent rise in pressure in the right atrium. In case I.N. 451026, the cyanosis was clearly of the peripheral type, owing to the low cardiac output. The arteriovenou[illegible] oxygen difference was large.

ANGIOCARDIOGRAPHY

When typical clinical features and [illegible] characteristic roentgenologic picture are present, the indications for angiocardiographic examination in this condition are only relative. The anatomic and functional

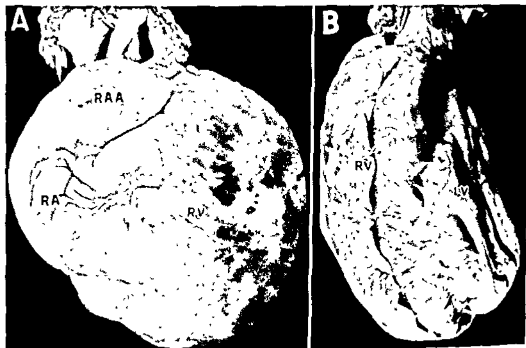


Fig. 629b.—Same case as in Figure 629a Right atrium and ventricle are enormously en- [illegible] extending like a large cap [illegible] per- [illegible] nu- [illegible] lus fibrosus) is greatly dilated (upper and right arrows in *D*) Cusps of the tricuspid [illegible] are greatly deformed, the medial and posterior cusps are fused into a funnel with a single, large opening at the bottom (*1* in *D* and *E*) *Margins of the cusps* are thickened, and a network of strands extends toward the walls of the right ventricle (*E*) The greater part of the anterior cusp is free. This results in two communications between the right atrium and ventricle (*1* and *2* in *D–F*) After severing of the fused part between the medial and posterior cusps, the three cusps together form a wide tube (*G*), which is attached to the ventricular wall just over *1 cm* below the annulus fibrosus In *H*, the heads of the pins mark the lower margin of the annulus fibrosus, arrow points to the marginal attachment of the cusps to the muscle wall *The chordae tendineae* are small and some are lacking Structure of the right ventricle is completely altered. Actual papillary muscles are lacking The thick muscular ridges, seen in the left region of the right ventricle (*M* in *E*), are probably abnormal papillary muscles The trabeculae are greatly *hypertro*phied and their course is entirely abnormal No thickening of the *actual* wall musculature [illegible] Pulmonary artery is narrow, the valve normal [illegible] tum (left lower arrow in *D*). *AF*, annulus fibro[illegible] rior cusps of tricuspid valve, *FoO*, fossa ovalis [illegible] right atrium, *RAA*, right auricular appendage, *SVC*, superior vena cava; *RV*, right ventricle; *LV*, left ventricle (*continued*).

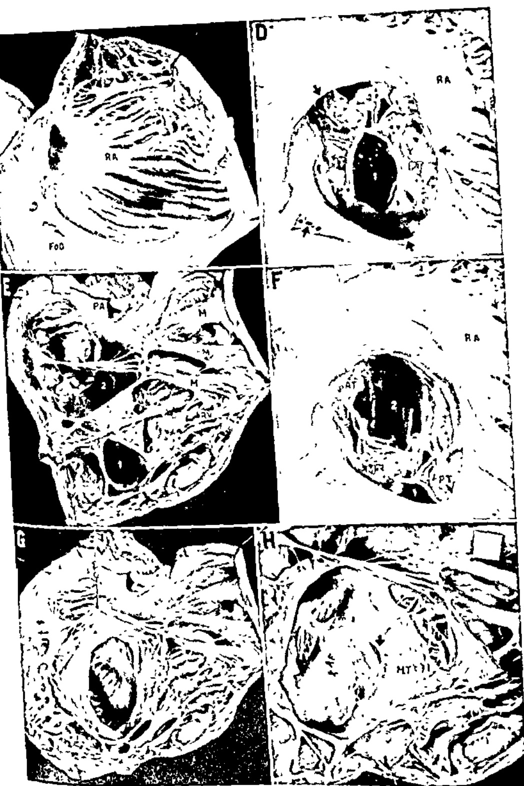


Fig 629b (cont)

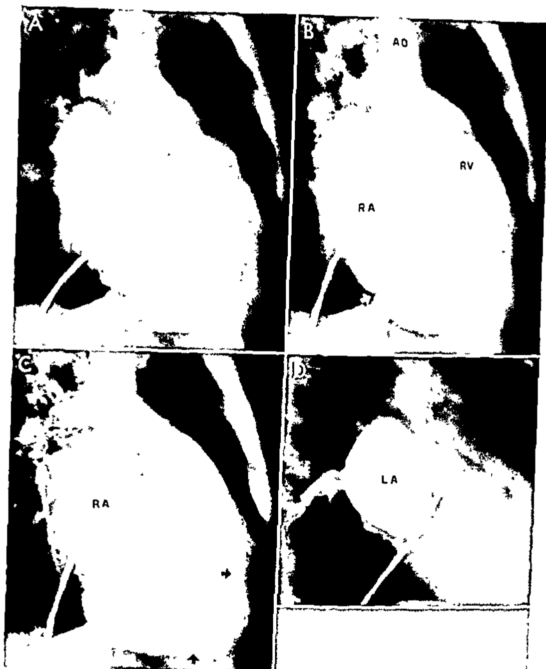


Fig. 630.—Ebstein's disease. Boy, aged 11 (B C 440203) An indentation in the outline (arrow in *B*) marks the border between the large right atrium (*RA*) and the right ventricle (*RV*) The tricuspid valve originates far down in the ventricle (lower arrow in *C*). The right atrium and the portion of the ventricle incorporated with it are seen in *A*, here, the contrast medium has reached the valvular plane In *C*, the sinus region of the ventricle is also opacified, so that both sides of the tricuspid valve are outlined (cf. *E*) and the valve (upper arrow in *C*) is distinctly visualized Left atrium (*LA*) small *AO*, aorta



Fig 631.—Ebstein's ...) Contrast medium injected into ... l regurgitation of contrast medium ... (*RA*) The tricuspid valve is situ- ... ılar plane, arrow in *F* points to the ... ving to reduced flow *A–D*, lateral ... ...ntal view

data that can be provided by angiocardiography are of limited value, particularly since surgical correction of the tricuspid anomaly has not hitherto been successful. Moreover, it has been questioned whether angiocardiography is not associated with greater risks in this condition that in most other forms of heart disease. Several deaths have been reported in direct connection with this examination.

A typical angiocardiographic picture, in which the right atrial enlargement, as well as the abnormal site of the tricuspid valve and the "atrialized" portion of the right ventricle appear, was described in 1951 by Soloff *et al.* (617). Similar observations have been made by other authors (68, 393, 459). These features are not, however, constant. Their depiction is hampered by dilution of the contrast medium in the usually greatly dilated right atrium, regurgitation in the tricuspid orifice—which further decreases opacification—and the inappreciable thickness of the tricuspid valve. When there is a right to left shunt, the left side of the heart is depicted simultaneously with the right atrium and ventricle, so that the details of the tricuspid region become still more difficult to identify.

In three of our cases the contrast medium was injected into the right atrium The enormous enlargement of this chamber in one of them is seen in Figure 629a. In the right ventricle, there was marked dilatation of the infundibulum and the sinus region. Displacement of the tricuspid valve could be established in all three cases, but the "atrialized" portion of the ventricle could not be delimited. It is seen in Figure 630 that on emptying of the right atrium the infundibulum of the right ventricle and the pulmonary artery are opacified before the sinus region of the ventricle; this is to be ascribed to the abnormal arrangement of the tricuspid leaflets A right to left shunt on the atrial level was present in one of these cases. No noteworthy cardiac arrhythmias occurred on injection of the contrast medium or after it.

In the fourth case, the contrast medium was injected into the right ventricle, in order to depict it selectively and to demonstrate directly the regurgitation in the tricuspid valve. The enlargement of the right ventricle and displacement of the tricuspid valve are shown in Figure 631. The massive leakage into the right atrium is considerably greater than can be explained by the presence of the catheter in the orifice, and presumably is due to the incompetent, abnormally situated tricuspid valve. No disturbances in heart rhythm were produced by introduction of the catheter into the ventricle. Injection of contrast medium into the right ventricle is warranted only when this applies In this case, injection caused only a few extrasystoles. Slow emptying of the right atrium was a feature common to all four cases. The pulmonary artery was well visualized in the two relatively mild cases and was only sparsely opacified in the other two

23 Tricuspid Atresia

IN TRICUSPID atresia, all venous blood passes from the right atrium to the left atrium, usually through a patent foramen ovale. A large atrial septal defect is sometimes present. The arterial and the venous blood are completely mixed in the heart and, from the functional standpoint, there is a cor triloculare biatriatum. The right atrium is dilated and hypertrophied, owing to difficulties in emptying. The foramen ovale is seldom as wide as a normal tricuspid orifice. The left atrium and left ventricle are also enlarged, and the mitral orifice becomes dilated on account of the large blood flow. The right ventricle, in contrast, is small, and Taussig (650) has therefore denoted these cases as underdeveloped right ventricle. It was this feature that she regarded as characteristic of the roentgenologic appearance and the electrocardiogram.

The pulmonary circulation is usually maintained by the pulmonary artery, which in most cases takes its origin from the underdeveloped right ventricle, this, in turn, receives blood through a ventricular septal defect. The pulmonary flow is generally decreased, either because of pulmonary stenosis or owing to the fact that the ventricular septal defect is so small that it impedes the flow. Pulmonary atresia is sometimes present, in this event, the lungs receive blood from the aorta, through a patent ductus or collaterals. The right ventricle is then a blind pouch which communicates only with the left ventricle or comprises a separate, entirely closed cavity which may be so small that it is easily overlooked at autopsy. Microscopic examination may be required to demonstrate its existence (216).

Tricuspid atresia may be combined with transposition of the great vessels. This is of no consequence from the functional viewpoint, since the venous and the arterial blood are completely mixed. Astley *et al* (26) were unable to note any difference between the findings in cases with and without transposition. The factor of essential importance is the distribution of the blood between the pulmonary and the systemic circulation. Pulmonary stenosis may be present irrespective of whether the great vessels are at the normal site or are transposed.

Our series contains 14 cases of tricuspid atresia. In four, the diagnosis was verified at autopsy. Cardiac catheterization and angiocardiography were performed in 10 cases, and venous angiocardiography alone, in an additional two cases.

CLINICAL FEATURES

The age distribution, anatomy of the malformation, and most important signs, symptoms, and findings are recorded in Table 21.

The *symptoms* vary according to the size of the pulmonary flow. One of our patients

TABLE 21.—TRICUSPID ATRESIA: ANATOMY OF MALFORMATIONS, SIGNS, SYMPTOMS, AND CLINICAL FEATURES IN 14 CASES

CASE	OTHER MALFORMATIONS*	GENERAL CONDITION	CYANOSIS	LIVER ENLARGEMENT	MURMUR	VASCULARITY OF LUNGS (X-RAY EXAM.)
Y.W. 540101 (girl, 2 wk.)	*Autopsy*: Pulm. atresia + underdev RV + patent foramen ovale + pat. ductus arteriosus	*Sudden death at age 2 mo*	*Severe since birth*	0	0	Decreased
S.L. 530307 (boy, 7 *mo*)	*Autopsy*: Pulm. atresia + underdev. RV + VSD + ASD + patent ductus arteriosus + mirror-image dextrocardia	Relatively good development; acute exacerbation with infection, death at age 1 yr	Severe since birth	0	0	Decreased
M.C. 520907 (girl, 2 mo.)	*Autopsy* + *Angio*: Patent *foramen ovale* + small VSD + underdev. RV	Attacks of cyanosis, unconsciousness; death at age 3 mo.	Severe since birth	Slight, no pulsations	Faint systolic	Decreased
G.H. 490707 (girl, 3⅓ yr.)	*Cath.* + *Angio*: Pulm atresia† + ASD + pulm circ. via bronchial arteries†	Walks only about 10 m	Severe since birth	Moderate; pulsating	Harsh systolic (grade 4) 2nd R I S	Decreased
P.C. 500324 (boy, 3⅗ yr.)	*Cath.* + *Angio*: ASD + wide VSD + underdev. RV + pulm stenosis + transpos. of aorta	Walks only 100 m	Severe since birth	0	Harsh systolic (grade 4) 2nd L I S.	Decreased
A.E. 530130 (girl, 1 yr 1 *mo*)	*Cath.* + *Angio*: ASD + wide VSD + underdev RV; no pulm. stenosis	Poor weight gain; gen'l condition good	Slight on exertion	0	Harsh systolic (grade 4) 3rd L I S	Normal
H.M. 460212 (boy, 6¼ yr.)	*Angio*: ASD + transpos of PA and aorta, both take origin from LV + no RV identified	Walks over 1,000 m	Moderate since birth	Slight; no pulsations	Harsh systolic 3rd R I S	Slightly increased
I.K. 531109 (girl, 7 mo.)	*Cath.* + *Angio* + *Autopsy*: Patent foramen ovale + small VSD + underdev RV	Attacks of cyanosis, death postop at age 7 mo	Severe since age 5 mo	Moderate; no pulsations	Harsh systolic apex	Decreased
H.K. 530209 (boy, 1 mo.)	*Cath* + *Angio*: ASD + VSD + transpos of PA and aorta + pulm stenosis	Attacks of cyanosis, tube feeding, death at age 5 mo	Severe since birth	Moderate, no pulsations	Faint systolic	Decreased

*Cath. = cardiac catheterization. Angio = angiocardiography.
†Verified at operation.

urvived only two months, whereas another was alive at 13 years of age and was only moderately disabled. Patients have been known to reach adult age (108). The arterial oxygen saturation is decreased, but with a large pulmonary flow, it is not invariably so severe that cyanosis develops. Two of our patients exhibited only slight cyanosis The majority nevertheless showed marked cyanosis from birth

The *physical signs* are not characteristic. A precordial bulge was present in none of our cases There may be no murmur whatever, but a systolic murmur is often heard and may be very loud It has no typical localization Both a ventricular septal defect and pulmonary stenosis may be responsible for the murmur. The second sound is usually pure, since the pulmonary component is entirely lacking (in pulmonary atresia) or is so faint that it cannot be detected (in the presence of decreased pulmonary flow). In one case (A.E. 530130) with normal vascularity of the lungs, both the aortic and the pulmonary components of the second sound nevertheless appeared over the pulmonary area on the phonocardiogram

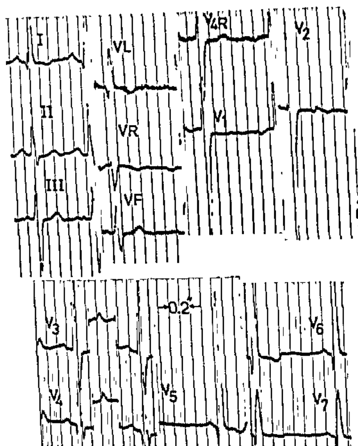


Fig 632.—Electrocardiogram in tricuspid atresia Boy, aged 6 (H M 460212). Transposition of pulmonary artery and aorta, but both take their origin from the left ventricle Right ventricle could not be identified

When the atrial septal defect or the patent foramen ovale is small, the right atrium may have difficulty in emptying and therefore becomes dilated and hypertrophic During atrial systole, the rise in pressure is extremely marked and causes

symptoms in the form of a pathologic venous pulse and a palpable hepatic pulse. If, on the contrary, the right atrium can be drained without difficulty through a large atrial septal defect, these symptoms are lacking (650). The liver was enlarged in five of our cases.

Electrocardiography

Left ventricular hypertrophy demonstrable on the ECG in a cyanotic child is characteristic of tricuspid atresia. The underdeveloped right ventricle and the dilatation and hypertrophy of the left ventricle and atrium and, in particular, of the right atrium cause characteristic changes in the electrocardiogram.

In our cases, there was left axis deviation. The chest leads were recorded in all but two cases. In V_{4R}, the R wave was low and the S wave deep. In V_{6-7}, the R wave was tall and the S wave small or lacking. The P wave was tall over the right precordium and tall, peaked and notched in leads I and II. The S-T interval was depressed in three cases, and in two of them the T wave was inverted (Fig. 632). A left bundle-branch block was recorded in one case. A rotation anomaly was present in two cases, there was right axis deviation and the chest leads showed a mirror image as compared with the other cases.

ROENTGENOLOGIC EXAMINATION

Opinions have varied with respect to the possibilities of diagnosing tricuspid atresia on roentgenologic examination. Taussig (650) has stated that the configuration is so typical that underdevelopment or absence of the right ventricle can be established. Wittenborg *et al* (708) found that there were, as a rule, no specific roentgenologic features. They expressed the view that a differentiation from tetralogy of Fallot cannot be made in the majority of cases, since it is not possible to determine on the basis of the roentgenogram whether the right or the left ventricle is enlarged Astley *et al.* (26) raised the objection that, in tricuspid atresia, the shape of the ventricular area is characteristic, with angulation and local prominence of the left superior cardiac border in its lateral and dorsolateral aspects. They ascribed this feature to overlapping of an enlarged left auricular appendage and stated that it was specific for tricuspid atresia.

We made a roentgenologic examination in 14 cases of tricuspid atresia: 10 without and four with transposition of the great vessels.

In our series, an enlarged ventricular area of the typical shape described by Astley *et al.* was distinctly visible in six cases, but was never pronounced (Figs. 633 and 634). It was nevertheless established by angiocardiography and autopsy that this part of the heart belonged to the left ventricle, whereas the left auricular appendage formed the superior, medial part of the cardiac border. The harmonious curvature of the lateral segment of the left cardiac border can be attributed entirely to the left ventricle. The same typical shape of the left cardiac border has, however, been observed in other forms of congenital heart disease as well (cf. Fig. 262, p. 281).

Underdevelopment of the right ventricle can seldom be demonstrated directly on roentgenologic examination. Only in one case did the right anterior segment of the cardiac border show such a distinct concavity that it could be ascribed to hypoplasia of the right ventricle (Fig. 635). The absence of a bulging ventricular outline below the aorta cannot be taken as evidence of underdevelopment of the right ventricle, since the same shape may be seen in tetralogy of Fallot, owing to the anterior position of the aorta. We found that the lower, anterior segment of the heart was often strikingly rounded in the lateral projection. This is presumably because it is formed by the left ventricle. No increase in contiguity to the thoracic wall was seen in our cases.

Unquestionable enlargement of the right atrium could be established in only four cases, in which the atrium exhibited a greatly elongated and bulging border (Fig. 636) At angiocardiography or autopsy, the







Fig. [illegible]
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identified. Greatly reduced vascularity of lungs, vascular markings have a reticular appearance, probably representing a collateral circulation

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mentary right ventricle lies anteriorly, slightly to the left of the atrioventricular border. Atresia of the pulmonary artery (Fig 39, p 39) Long, fairly wide patent ductus *FO*, foramen ovale, *LA* and *RA*, left and right atria, *LV*, left ventricle, *MV*, mitral valve, *PM*, papillary muscles.

atrium was found to be enlarged in every case. Thus, this observation applied even in the presence of the distinct concavity in the anterior and lateral border of the right segment of the heart described as characteristic of tricuspid atresia (Fig. 635). It may therefore be inferred that the possibilities of evaluating the size of the right atrium by ordinary roentgenologic examination are limited One of the main reasons is that the position of the atrium is more or less abnormal, owing to underdevelopment of the right ventricle (708)

Snow (614) has pointed out that, in oblique projections, the pulsations over the anterior and posterior areas of the heart are asynchronous, owing to increased systolic activity of the right atrium. Their appearance even in an extremely oblique projection seems to be specific for tricuspid atresia The same phenomenon can, however, be observed in other forms of heart disease with an enlarged, hypertrophied right atrium if the examination is made in a less extreme oblique projection

The pulmonary artery could be identified in only three of the cases in which the great vessels were not transposed. The vascularity was usually sparse, and in some cases the vascular markings had the same appearance as in a collateral circulation (Fig 633). An increased blood volume in the lungs occurred in only one case. This was one of the four in which the great vessels were transposed. In the remaining three cases, the vascularity was normal or reduced



Fig. 634a.—Tricuspid atresia, patent foramen ovale, rudimentary right ventricle with closed ventricular septum, pulmonary atresia, and patent ductus arteriosus. Girl, aged 14 days (Y.W. 540101). see Figure 39, p. 39. Left ventricle is grossly enlarged, and left border, which is greatly curved, has typical shape Medial, superior part of left border of the heart is formed by the enlarged left auricular appendage (arrow in *A*) Main trunk of the pulmonary artery not visible, considerable reduction in vascularity of lungs.

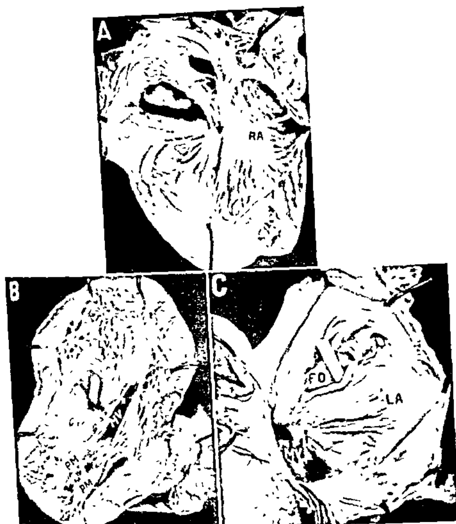
The left atrium was slightly or moderately enlarged in nine cases.

The width of the aorta was hard to judge, particularly in the infants. Unquestionable dilatation could only rarely be shown. A right-sided aortic arch, which has been stated to be about as common as in tetralogy of Fallot (708), was observed in two cases (Figs 635 and 637).

The heart volume was increased in every case In the majority the enlargement was moderate, and only exceptionally was it more pronounced.

Dextrocardia was present in two cases

From the roentgenologic viewpoint, it may be difficult to distinguish between tricuspid atresia and tetralogy of Fallot, es-



Fi [illegible] ... [illegible] right atrium is a small [illegible] passe[illegible] the la[illegible] ...mentary right ventricle lies anteriorly, slightly to the left of the atrioventricular border Atresia of the pulmonary artery (Fig 39, p 39) Long, fairly wide patent ductus FO, foramen ovale, LA and RA, left and right atria, LV, left ventricle, MV, mitral valve, PM, papillary muscles

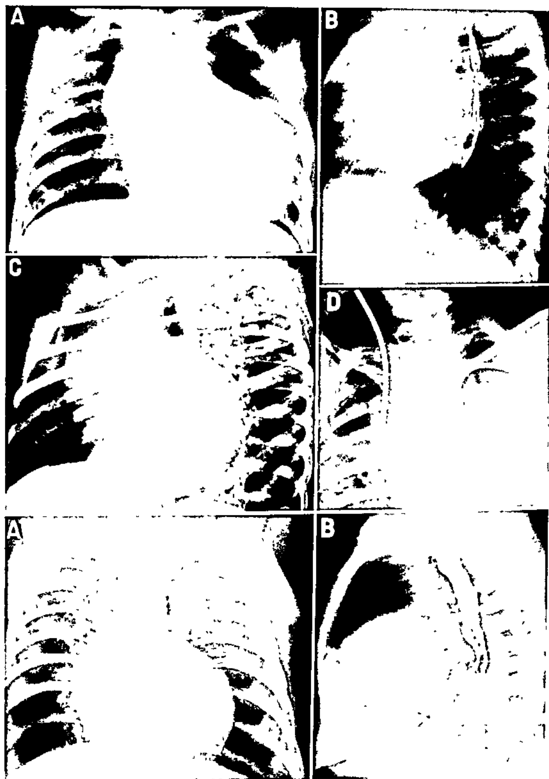


Fig. 635 (*above*) —Tricuspid atresia, pulmonary atresia, and atrial septal defect Girl, aged 3 [illegible] ht atrium cannot be esti- [illegible] border (arrow in *A*) is [illegible] arch The [illegible] Greatly re- [illegible]

[illegible] icular septal defect, and

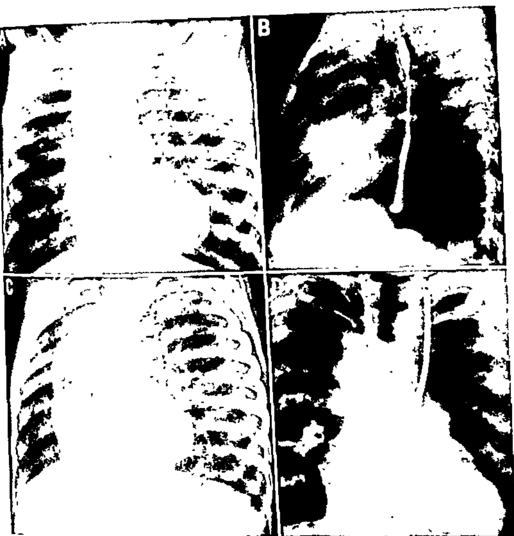


Fig 63̄ ... superior v... ance as in ...

...ings Catheter inserted in left superior vena cava

small right ventricle Girl, aged 7 months (I-L K 531109), see Figure 642 As in the preceding cases considerable enlargement of left ventricle, dilatation of right and left atria as well, pronounced reduction in vascularity of lungs

pecially in combination with pulmonary atresia (Fig. 247, p. 268). The shape of the heart presents great similarities, its size is approximately the same, and decreased vascularity of the lungs is typical of both conditions. Unquestionable differences are nevertheless sometimes present. In tricuspid atresia, the apex—which may be round or blunted—is never angulated, a feature which is often characteristic of tetralogy of Fallot (Fig. 240, p. 262). Moreover, the lateral and superior segments of the ventricle differ in appearance. Despite the decreased vascularity of the lungs, the left atrium often bulges toward the esophagus in tricuspid atresia. In tetralogy of Fallot, dilatation of the left atrium is present only in rare cases with increased pulmonary blood volume. In many cases the similarities in the appearance are, however, so great and the differences so indistinct that it is not possible to make a roentgenologic differential diagnosis. This applied in our cases when tricuspid atresia was combined with dextrocardia (Fig. 638) or with transposition of the great vessels without an increase in vascularity of the lungs, or when the right ventricle was relatively large (Fig 637). In cases combined with rotation, the appearance may be entirely uncharacteristic (Figs. 639 and 640).

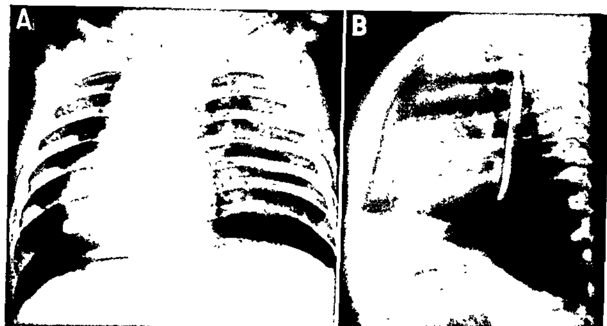


Fig. 638a.—Tricuspid atresia, ventricular septal defect, pulmonary atresia, rudimentary right ventricle, patent ductus arteriosus and dextrocardia. Boy, aged 6 months (S.L. 530307). Appearance as in tetralogy of Fallot Reduced vascularity of lungs.

CARDIAC CATHETERIZATION

When the catheter is directed toward the tricuspid orifice, it cannot be advanced into

Fig. 638b.—Same case as in Figure 638a A small, shallow dimple (arrow in *A*) is seen at the site of the tricuspid orifice A large valve covers a wide foramen ovale (probe in *A* and *C*). Right atrium is large and trabeculae are hypertrophied. Left atrium is of normal size; mitral orifice is wide. Small right ventricle with completely abnormal anatomy, situated anteriorly to the left. Atresia immediately below the pulmonary valve (arrow in *D*). Large ventricular septal defect, through which papillary muscles run into a large left ventricle. Right-sided aortic arch. Pulmonary artery is narrow and is patent as far as the valvular plane. Patent ductus is severed *AO*, aorta; *IVC* and *SVC*, inferior and superior venae cavae, *LA* and *RA*, left and right atria, *LV* and *RV*, left and right ventricles, *MV*, mitral valve, *PA*, pulmonary artery, *VSD*, ventricular septal defect. ⟶



Fig. 638b (legend on facing page)

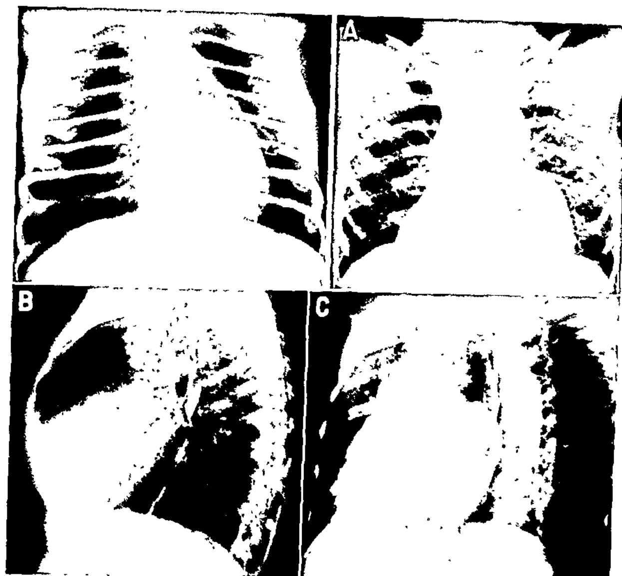


Fig. 639 (*above left*).—Tricuspid atresia, probably single ventricle, pulmonary stenosis, and rotation anomaly. Boy, aged 4 weeks (H K 530209), see Figure 647. Notch in left border (arrow) marks site of the aortic orifice. The ascending aorta is on the left side Greatly reduced vascularity of lungs.

Fig. 640 (*above right and below*).—Tricuspid atresia, single ventricle and rudimentary outlet chamber, pulmonary stenosis, and anomalous drainage of systemic veins Boy, aged 8 (H M 451003), see Figure 650. Great increase in heart volume, considerable enlargement of left atrium and the anteriorly situated ventricle. Bulge in the superior part of the left cardiac border corresponds to the infundibulum of an outlet chamber The greatly expanded right cardiac border is due to a pouch-like bulge, probably the right atrium Great dilatation of aorta Main trunk of the pulmonary artery cannot be identified. Reduced vascularity of lungs, suggested collateral circulation.

the right ventricle, for it slips upward and to the left and passes into the left atrium. The large shunt from the right to the left atrium can be demonstrated by gas analysis. These are the most characteristic findings on cardiac catheterization in tricuspid atresia.

The same findings may, however, be noted in severe tricuspid stenosis. Angiocardiography is required for a detailed anatomic analysis.

In nine of our 10 cases in which cardiac catheterization was performed, the catheter could be passed into the left side of the heart (Table 22). A large right to left interatrial shunt was present in every instance. In one case there was a single atrium from the functional aspect. Systemic veins opened into the left atrium. The right atrium was rudimentary, forming an outpouching on the right side of the heart, and was in wide communication with the left atrium (Fig. 650).

In five cases, the mean pressure in the right atrium was close to the upper limit for the normal range of variations, a tall *a* wave was sometimes present.

TABLE 22.—TRICUSPID ATRESIA: MOST IMPORTANT FINDINGS ON CARDIAC CATHETERIZATION IN 10 CASES*

Case	O_2 Content, vol. %						O_2 Cap., vol. %	Pressure, mm Hg RA	
	SVC	IVC	RA	LA	PV	LV		Syst	Mean
G.R. 490707	11.0	11.4	11.4	—	—	—	21.5	10	5
P.C. 500324	19.3	19.6	20.6	23.3	30.6	22.4	31.2	8	5
A.E. 530130	8.7	8.9	9.2	10.6	—	10.8	17.4	4	0
I.K. 531109	8.4	8.2	8.2	9.6	17.0	—	17.5	8	5
H.K. 530209	4.3	5.0	4.8	—	12.2	5.8	12.3	2	0
H.M. 451003	15.5	14.8	17.9†	18.9‡	30.0	19.2	30.9	9	5
A.F. 530307	10.0	11.2	10.1	11.2	19.8	12.4	21.2	11	8
K.H. 481217	17.8	17.8	18.1	24.4	—	23.2	28.7	7	4
P.P. 520411	13.8	13.4	13.2	17.5	21.0	15.3	21.4	4	3
J.E. 430429	13.6	12.8	13.6	—	19.8	15.2	20.8	6	3

*For abbreviations see Table 1, p. 119 †At orifice of SVC (single atrium) ‡At orifice of pulmonary vein

ANGIOCARDIOGRAPHY

The essential indication for angiocardiography is to demonstrate the existence of tricuspid atresia and, if possible, to visualize the pulmonary artery.

Tricuspid atresia, as well as the communication between the atria, can be established by means of intravenous injection of contrast medium or of injection into the right atrium. It is usually possible to obtain satisfactory visualization of the left atrium and left ventricle. The right ventricle, on the other hand, which is the last of the cavities to become filled, can be distinguished from the other chambers of the heart only incompletely or not at all. The pulmonary artery is generally clearly visible only when it is normal in size or dilated. A patent ductus is occasionally visualized. Complementary injection of the contrast medium into the left ventricle is, as a rule, necessary for more consistent visualization of the right ventricle and the pulmonary artery. Consequently, several angiocardiographic examinations are required for a detailed anatomic analysis in tricuspid atresia. On practical grounds, such a special investigation is warranted only when the pulmonary artery is poorly visualized and an anastomotic operation is contemplated.

Angiocardiographic examination was made in 12 of our cases. In two the injection was made intravenously, in six into the right atrium, in one into the left atrium, and in two into the left ventricle. In one case two investigations were performed; the contrast medium was injected in one into the atrium and in the other into the ventricle. Frontal and lateral projections

were used. The fact that the atrioventricular plane is not depicted optimally in these projections is of minor importance. This is because of lack of filling of the right ventricle in the first exposures in the series shows that tricuspid atresia is present. It is of greater consequence from the surgical point of view that the position of the pulmonary artery in relation to both the aorta and the systemic arteries is established best in the frontal and lateral projections.

moderately enlarged in several cases. In all but one, the appendage was situated to the left, beside the vessel trunk. Janker and Hallerbach (357) have stated that the left atrium lies farther to the left than normally, and have attributed this shift to rotation, on the grounds of enlargement of the left ventricle. The atrium was at the normal site in our series.

Enlargement of the left ventricle, which was a constant feature, was shown on

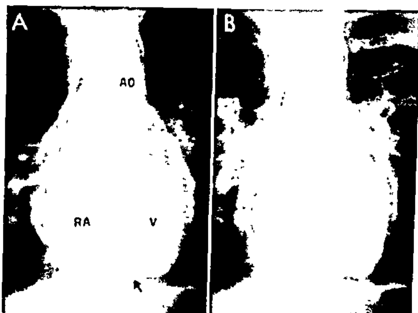


Fig. 641.—Tricuspid atresia, single ventricle, transposition of great vessels, and pulmonary stenosis Boy, aged 3 (P.P 520411) Right atrium is greatly enlarged Its medial border (arrow in *A*) is distinct and lies far to the left, partly occupying the site of the right ventricle Aorta and pulmonary artery are transposed, they take their origin from a common ventricle (*V*).

The right atrium, including the appendage, was enlarged in every case. The borderline between the atrium and the right ventricle was sharply defined and was often strikingly medial in position (Fig 641). In one case the auricular appendage lay to the left of the vessel trunk and partly behind it.

The left atrium was filled via the interatrial communication. Its width can be determined only if the septum is depicted tangentially; this seems to necessitate the use of oblique projections In the frontal view, the atria probably show some degree of overlapping, so that the communication appears to be wider than it actually is The left atrium, as well as the appendage, was

angiocardiography to be due to both dilatation and hypertrophy.

The right ventricle was visualized clearly in only three cases. It was relatively large (Figs. 645 and 646) and had a wide communication with the left ventricle. In the two autopsy cases, in which the investigation had included angiocardiography but the right ventricle could not be identified, or only partially, by this means, the septal defect was found to be extremely narrow (Fig. 642) Experience in other series (159) has also shown that the right ventricle is hard to identify. A typical finding is that no filling takes place in that segment of the heart which normally corresponds to part of the

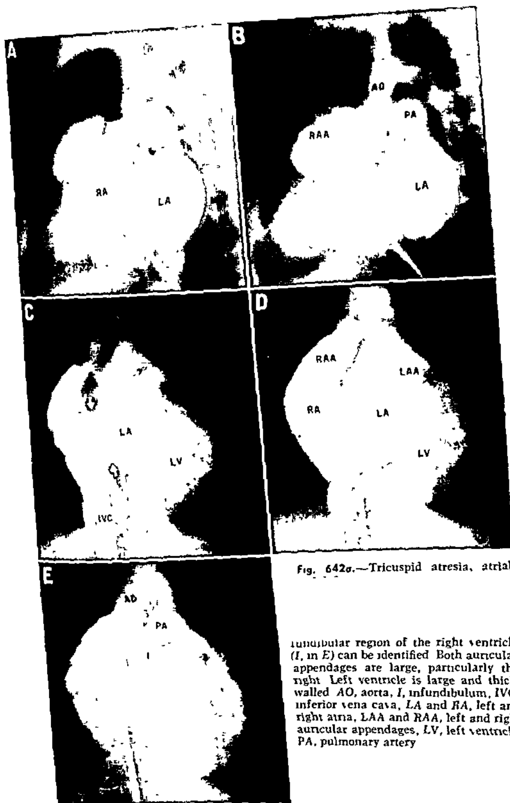


Fig. 642a.—Tricuspid atresia, atrial

infundibular region of the right ventricle (*I*, in *E*) can be identified Both auricular appendages are large, particularly the right Left ventricle is large and thick-walled *AO*, aorta, *I*, infundibulum, *IVC*, inferior vena cava, *LA* and *RA*, left and right atria, LAA and RAA, left and right auricular appendages, *LV*, left ventricle, PA, pulmonary artery

were used. The fact that the atrioventricular plane is not depicted optimally in these projections is of minor importance. This is because of lack of filling of the right ventricle in the first exposures in the series shows that tricuspid atresia is present It is of greater consequence from the surgical point of view that the position of the pulmonary artery in relation to both the aorta and the systemic arteries is established best in the frontal and lateral projections.

moderately enlarged in several cases In al[l] but one, the appendage was situated to th[e] left, beside the vessel trunk. Janker an[d] Hallerbach (357) *have stated that the lef[t]* atrium lies farther to the left than normally and have attributed this shift to rotation on the grounds of enlargement of the lef[t] ventricle. The atrium was at the normal sit[e] in our series.

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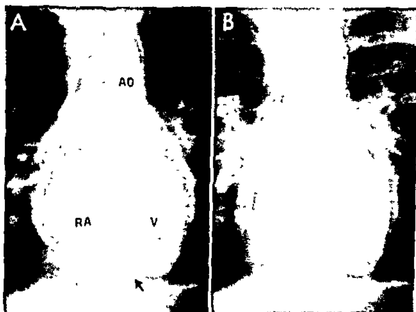


Fig. 641.—Tricuspid atresia, single ventricle, transposition of great vessels, and pulmonary stenosis. Boy, aged 3 (P.P. 520411) Right atrium is greatly enlarged. Its medial border (arrow in *A*) is distinct and lies far to the left, partly occupying the site of the right ventricle. Aorta and pulmonary artery are transposed, they take their origin from a common ventricle (*V*).

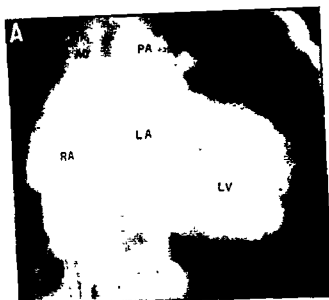
The right atrium, including the appendage, was enlarged in every case. The borderline between the atrium and the right ventricle was sharply defined and was often strikingly medial in position (Fig 641) In one case the auricular appendage lay to the *left of the vessel trunk and partly behind it.*

The left atrium was filled via the interatrial communication Its width can be determined only if the septum is depicted tangentially, this seems to necessitate the use of oblique projections. In the frontal view, the atria probably show some degree of overlapping, so that the communication appears to be wider than it actually is The left atrium, as well as the appendage, was

angiocardiography to be due to both dilatation and hypertrophy.

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only partially, by this means, the [illegible] fect was found to be extremely narrow (Fig 642). Experience in other series (159) has also shown that the right ventricle is hard to identify. A typical finding is that no filling takes place in that segment of the heart which normally corresponds to part of the



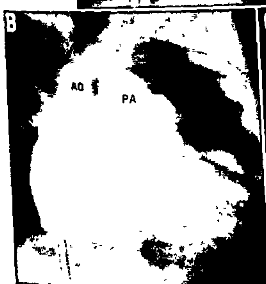
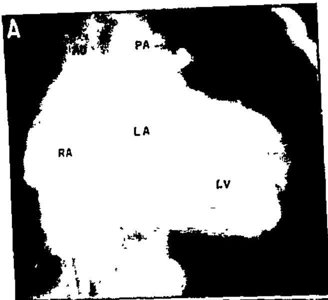


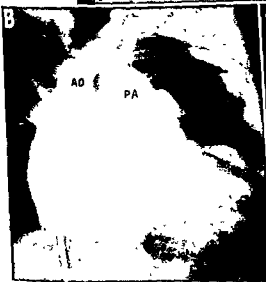


Fig. 682. [illegible]



Fig. 642b.—Same case as in Figure 642a Dilatation of both atria, greater of the right (arrow in *A*, the atresia, in *B*, mitral valve) Large atrial septal defect Small right ventricle (arrow in *C*, the slit-shaped sinus region) lies far to the right in the anterior wall, directly beside the atrioventricular border, it consists mainly of the infundibulum (*RV* in *D*) A 3 mm wide ventricular septal defect opens at the lower margin of the infundibulum (in *C* and *D*, the probe lies in the defect). Large left ventricle with great hypertrophy of trabeculae and wall muscles, narrow pulmonary artery with normal valves, wide aorta *AO*, aorta, *ASD*, atrial septal defect, *IVC*, inferior vena cava; *LA* and *RA*, left and right atria, *LV* and *RV*, left and right ventricles, *MV*, mitral valve, *PA*, pulmonary artery





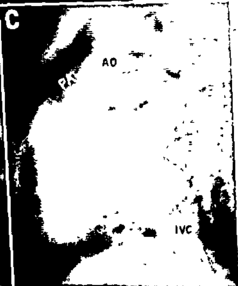


Fig 643.—Tricuspid atresia, atrial septal defect, ventricular septal defect, and small right ventricle Girl, aged 1 yr (A E 530130) The atrial septal defect is probably very wide Margins of the right ventricle cannot be identified *AO*, aorta, *IVC*, inferior vena cava, *LA* and *RA*, left and right atria, *LV*, left ventricle, *PA*, pulmonary artery.

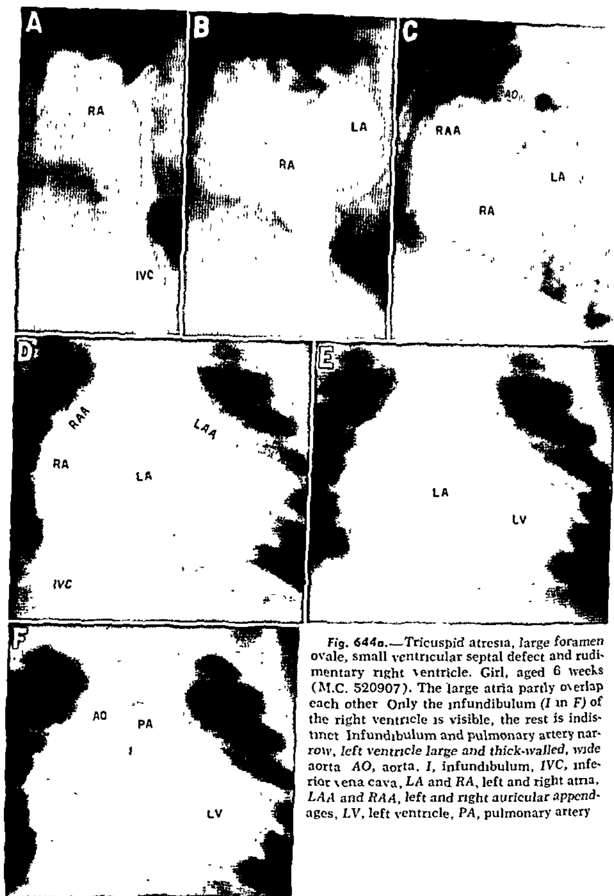


Fig. 644a.—Tricuspid atresia, large foramen ovale, small ventricular septal defect and rudimentary right ventricle. Girl, aged 6 weeks (M.C. 520907). The large atria partly overlap each other. Only the infundibulum (*I* in *F*) of the right ventricle is visible, the rest is indistinct. Infundibulum and pulmonary artery narrow, *left ventricle large and thick-walled*, wide aorta. *AO*, aorta, *I*, infundibulum, *IVC*, inferior vena cava, *LA* and *RA*, left and right atria, *LAA* and *RAA*, left and right auricular appendages, *LV*, left ventricle, *PA*, pulmonary artery



Fig. 644b. Same case as in Figure 644a Right atrium is large, with highly developed trabec- ... the site of tricuspid atresia (arrow in A). ...

FO, foramen ovale, LA
left and right ventricles,

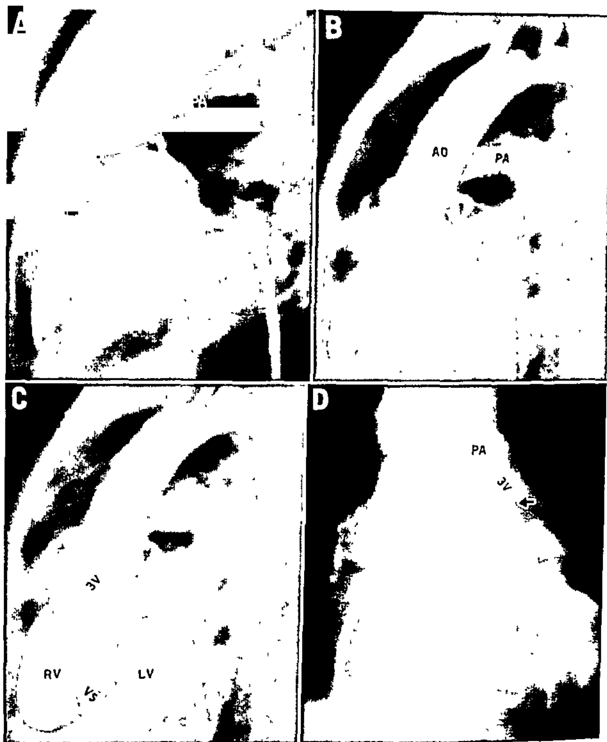


Fig. 645.—Tricuspid atresia, large ventricular septal defect, infundibular stenosis, left-sided superior vena cava and over-riding aorta Boy, aged 4 (P C 500324) Catheter is introduced through the mitral orifice and ventricular septal defect, tip lies in right ventricle Right ventricle is distinctly larger than in the preceding cases Considerable stenosis of the entire infundibulum, greatest at the level of the ostium [illegible] arch Degree of over-riding is difficult to judge, sinc[illegible] orta, *LV* and *RV*, left and right ventricles, *PA*, pulm[illegible] cular septum (*continued*).

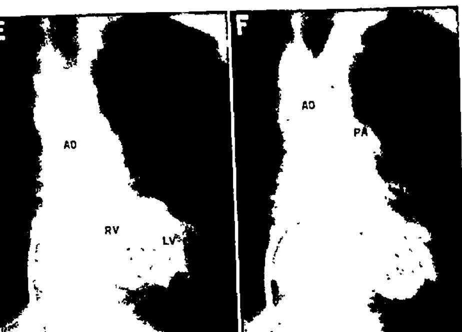


Fig 645 (*cont*)

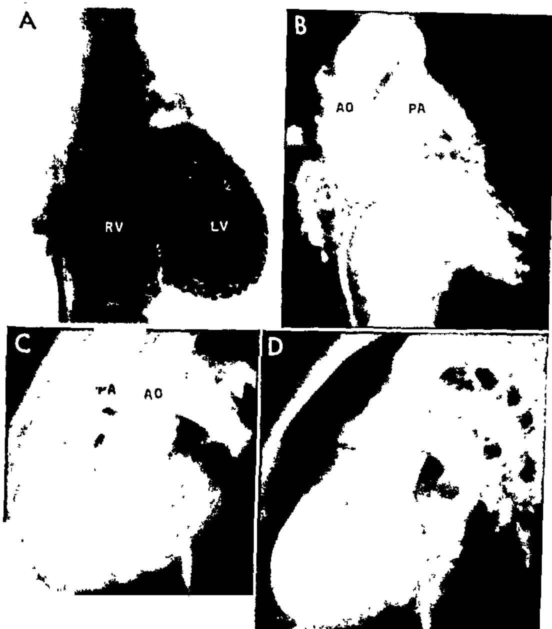
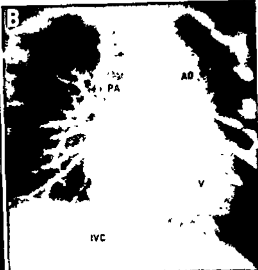


Fig. 646.—Tricuspid atresia Boy, aged 13 (J-A.E. 430429). Injection of contrast medium into left ventricle (*LV*) communicating with right ventricle (*RV*) through a wide ventricular septal defect. Reduced blood flow through pulmonary circuit Aorta (*AO*) increased in width. Large atrial septal defect but no demonstrable communication between right atrium and ventricle

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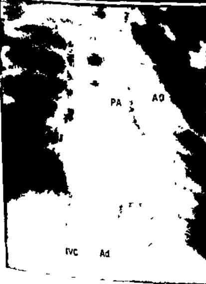
Fig. 647.—Tricuspid atresia, probably single ventricle, pulmonary stenosis, and corrected transposition of great vessels. Boy, aged 4 weeks (H K. 530209) Injection into inferior vena cava. Large reflux into pulmonary veins on both sides (*B*, *H* and *I*) Wide interatrial communication. There is probably a common ventricle with an outlet chamber to the aorta, which lies to the left. The outlet chamber is filled later than the rest of the ventricle (*A*). As far as the valvular plane, the pulmonary artery is narrow Position and nature of the stenosis cannot be determined. *Ad*, descending aorta, *AO*, aorta, *I*, outlet chamber, *IVC*, inferior vena cava, *LA* and *RA*, left and right atria, *PA*, pulmonary artery, *V*, ventricle ⟶



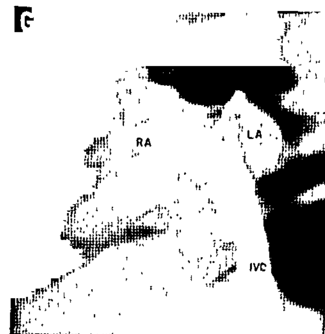

B
PA
AO
V
IVC


AO
I
V


D


PA
AO
IVC
Ad


F



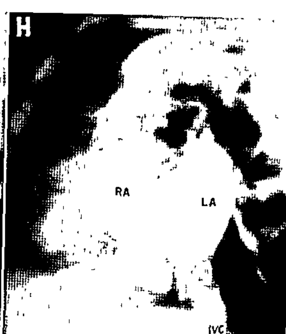






Fig. 647 *(cont)*

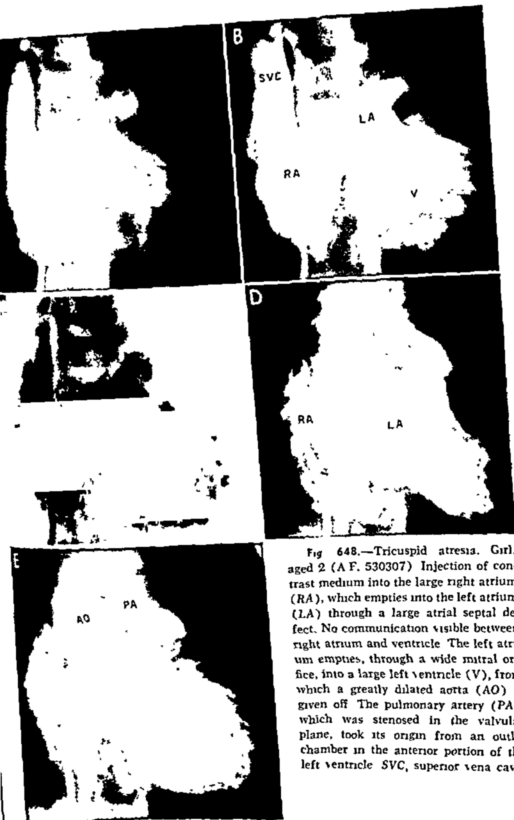


Fig 648.—Tricuspid atresia. Girl, aged 2 (A F. 530307) Injection of contrast medium into the large right atrium (*RA*), which empties into the left atrium (*LA*) through a large atrial septal defect. No communication visible between right atrium and ventricle The left atrium empties, through a wide mitral orifice, into a large left ventricle (*V*), from which a greatly dilated aorta (*AO*) is given off The pulmonary artery (*PA*), which was stenosed in the valvular plane, took its origin from an outlet chamber in the anterior portion of the left ventricle *SVC*, superior vena cava.



Fig. 649.—Tricuspid atresia. Boy, aged 6 (K-Y H 481217). Contrast medium injected into left atrium (*LA*), communicating with right ventricle (*V*), which is transposed. Aorta (*AO*) given off from right ventricle and rotated forward and to left Probable atresia of pulmonary artery at level of valve. Circulation in the pulmonary circuit is maintained chiefly through a wide patent ductus (*X*) and a moderately well-developed collateral circuit *G*, injection of contrast medium into inferior vena cava (*IVC*) at its entry into right atrium (*RA*) The atrium is large, has a dorsal position, and communicates through a high, posterior defect, about 1 cm wide, in the atrial septum (sinus venosus defect), with the left atrium, into which all its blood is emptied *PV*, pulmonary veins, *PA*, pulmonary artery (*continued*).

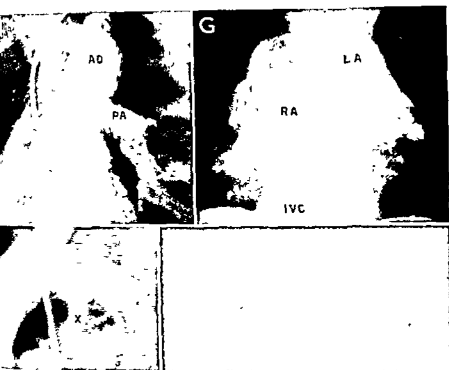


Fig. 649 (*cont*)

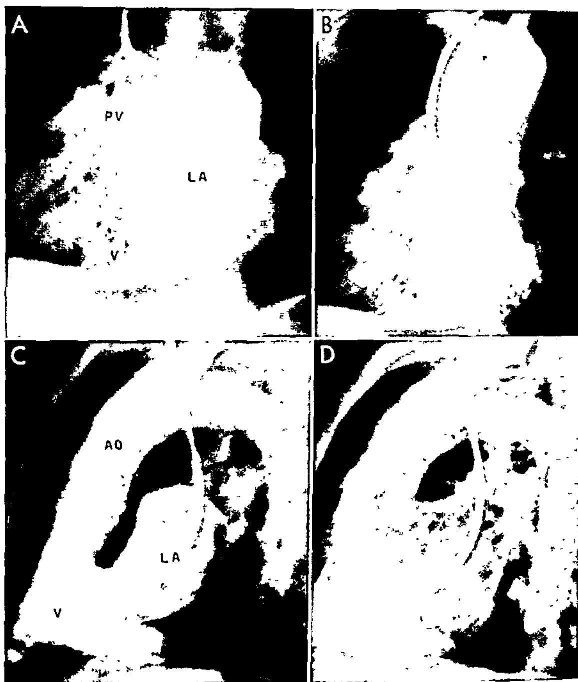


Fig. 649.—Tricuspid atresia Boy, aged 6 (K-Y.H. 481217). Contrast medium injected ıı left atrium (*LA*), communicating with right ventricle (*V*), which is transposed Aorta (*A* given off from right ventricle and rotated forward and to left Probable atresia of pulmon: artery at level of valve. Circulation in the pulmonary circuit is maintained chiefly througł wide patent ductus (*X*) and a moderately well-developed collateral circuit. *G*, injection of c(trast medium into inferior vena cava (*IVC*) at its entry into right atrium (*RA*). The atrium large, has a dorsal position, and communicates through a high, posterior defect, about 1 ı wide, in the atrial septum (sinus venosus defect), with the left atrium, into which all its blc is emptied. *PV*, pulmonary veins, *PA*, pulmonary artery (*continued*)

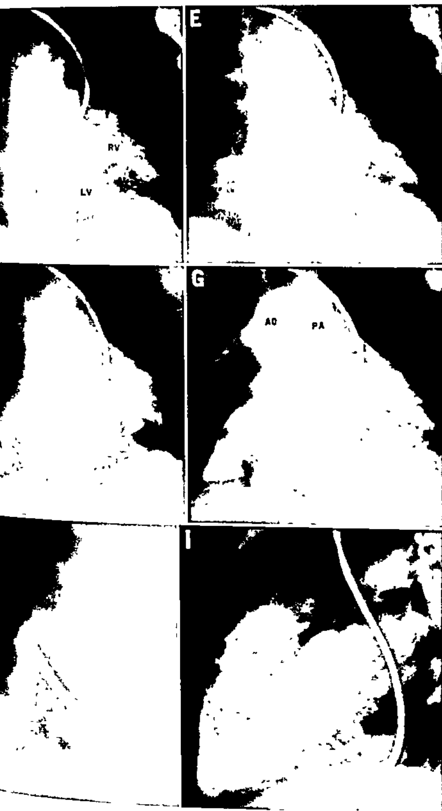


Fig. 650 (*cont*)



Fig. 650.—Tricuspid atresia, single ventricle and rudimentary outlet chamber, pulmonary stenosis and anomalous drainage of systemic veins. Boy, aged 8 (H M. 451003). Injection of contrast medium into *A–C*, atrium, *D–I*, ventricle. Inferior and left superior venae cavae open into a large atrium, lying dorsally and having the appearance of a left atrium In its right portion is an outpouching, probably representing a rudimentary right atrium or an auricular appendage The large atrium is drained into a large ventricle, lying farthest anteriorly. It is divided by a remnant of the septum into an underdeveloped right ventricle, lying on the left (*RV* and *I* in *D*) and a large portion from which the greatly dilated aorta originates (*LV* in *D*) Pulmonary artery ... inferior vena ... artery, ... *S*, ven-

24 Tricuspid Stenosis

[C]ONGENITAL TRICUSPID stenosis is rare [an]d is presumably always combined with [ot]her cardiac malformations The most [co]mmon associated anomalies are atrial [an]d ventricular septal defects Pulmonary [ste]nosis or pulmonary atresia is sometimes present and, occasionally, transposition of [t]he great vessels (584) According to Lev [(]421), the cusps may be short and thin, [w]ith small papillary muscles, or thickened, [w]ith stubby papillary muscles The cusps [a]re sometimes fused into a cone As a rule, [th]e foramen ovale is patent, and a varying proportion of the blood flow is shunted from the right atrium to the left. As is invariably the case in complicated anomalies, the classification is difficult It appears practical to base it on the malformation regarded as having the greatest functional importance. Tricuspid stenosis of a somewhat severe degree has such a profound effect on both the hemodynamics and the clinical features that one is justified in assembling in one group cases involving this anomaly.

The clinical features vary both with the

TABLE 23 —TRICUSPID STENOSIS· ANATOMY OF MALFORMATIONS, SIGNS, SYMPTOMS AND CLINICAL FINDINGS IN 3 CASES

[C]ASE	OTHER MALFORMATIONS*	GENERAL CONDITION	CYANOSIS	LIVER ENLARGEMENT	CARDIAC FINDINGS
[4]60728 ([..]r, [..] yr.)†	*Cath* + *Autopsy* Single ventricle + transpos of great vessels + pulm stenosis (4 × 4 mm) + patent foramen ovale (1 5 cm) + dextrocardia, tricuspid orifice, 4 × 4 mm	Can walk max, 25 m on level	Severe since birth	0	Acc 2nd sound over pulm area, syst murmur, 3rd L.I S
E.H. 430128 (girl, 11 yr)	*Cath* + *Angio* Valvular pulm stenosis with intact vent septum + small RV + patent foramen ovale‡	Can walk 1,000 m	Severe since birth	Slight, not pulsating	Loud systolic murmur, 3rd L.I S
L.E. 461207 (boy, 8 yr)	*Cath* + *Angio* ASD + valvular pulm stenosis + small but hypertrophic RV	Mod disabled	Slight since age 3 yr.	0	Systolic murmur grade 4, 2nd L.I S
D.B. 460508 (girl, 8 yr)	*Cath* + *Angio* Tetralogy of Fallot + patent foramen ovale	Can walk about 100 m, unable to run	Severe since birth	0	Pure 2nd sound over pulm, area, short, early-syst murmur, 3rd L.I S

*Cath = cardiac catheterization Angio = angiocardiography
†Died of a cerebral abscess at age 16
‡Verified at operation.

inflow tract of the right ventricle (136, 146) (*see* Figs. 642–644 and 648).

In two cases infundibular stenosis was demonstrated and filling of the pulmonary artery was satisfactory (Fig. 645). The pulmonary artery was visualized in an additional eight cases, but the anatomy of the pulmonary orifice could not be evaluated in four of them, since the right ventricle was not visible (Fig. 643). Distinct valvular stenosis was depicted in two cases, in one there was moderate obstruction of the orifice, and in another it was atresic, the pulmonary artery being opacified through a patent ductus arteriosus (Fig. 649). A narrow pulmonary artery was seen in one case with a single ventricle (Fig. 647).

Slight or moderate dilatation of the aorta was a regular feature.

One case differed in many respects from the others (Fig. 650). Two investigations were made, the first with injection into the left atrium and the second with injection into the ventricle. The left superior and the inferior venae cavae opened into a large atrium, which had the appearance of a left atrium. In its right portion was seen an outpouching, which we supposed to be either a right atrium or an appendage. The left atrium drained into a large ventricle. It was divided into two parts by a small remnant of the septum, one was large and lay anteriorly to the right, and the other was small and lay on the left. The greatly dilated aorta originated from the former and the narrow pulmonary artery from the latter. No right superior vena cava could be identified in these studies



Fig 651a —T



degree of stenosis of the tricuspid orifice and with the nature of the associated malformations. Severe stenosis combined with a *patent foramen ovale gives rise to the* same signs and symptoms as tricuspid atresia. The larger the communication between the atria, the more easily can emptying take place and the less likely are signs of right atrial hyperactivity to occur.

Our series contains four cases of tricuspid stenosis. The complete diagnosis, as well as the most important symptoms and signs, are recorded in Table 23. In one case, the diagnosis was established only at autopsy. In the other two the diagnosis was based on cardiac catheterization and angiocardiography.

CLINICAL FEATURES

All four patients were cyanotic and moderately to severely disabled.

The symptoms and signs were dominated entirely by an associated pulmonary stenosis. Both the murmur and the pulmonary component of the second sound nevertheless differed from those usually found in pulmonary stenosis, this feature will be discussed in connection with the hemodynamics. In case A.A. 380728, the second sound over the pulmonary area was accentuated, owing to transposition of the aorta. *The liver was enlarged in one case, but exhibited no pulsations.*

ELECTROCARDIOGRAPHY

Pronounced right ventricular hypertrophy was present in three cases, it was combined in two with pulmonary stenosis with an intact ventricular septum and in the other with tetralogy of Fallot. Case A.A. 380728, in which the ventricular septum was lacking entirely and there were also transposition of the great vessels, pulmonary stenosis, and dextrocardia, exhibited left ventricular hypertrophy and counterclockwise rotation of the heart *All of the* patients had abnormally tall P waves over the right precordium, and two had them in leads I and II as well They were then broad and notched, indicating enlargement of both the right and the left atrium.

ROENTGENOLOGIC EXAMINATION

As far as we are aware, no descriptions have been given of congenital tricuspid stenosis as an isolated lesion. Brown *et al.* (108) described a case combined with an ostium primum defect and a small defect in *the muscular part of the ventricular* septum. The features were essentially the same as in their cases of tricuspid atresia. In our material as well, associated malformations, as recorded in Table 23, were present in all four cases.

In the first case (Fig. 651), dextrocardia was present, and this made the interpretation more difficult. The salient features were reduced vascularity of the lungs, a narrow aorta, an ordinary pulmonary artery, undilated atria, and a ventricular region lacking any typical shape.

In the second case (Fig 652), the remarkable feature was that the enlarged right atrium was partly interposed between the sternum and the anterior surface of the heart. As a result, the whole heart was somewhat dorsally displaced We have otherwise observed this phenomenon, which is unusual, only when the right ventricle is underdeveloped (cf. Fig. 658). The shape *of the ventricular region was uncharacteristic. The pulmonary artery had the usual* appearance, whereas the vascularity of the lungs was markedly decreased.

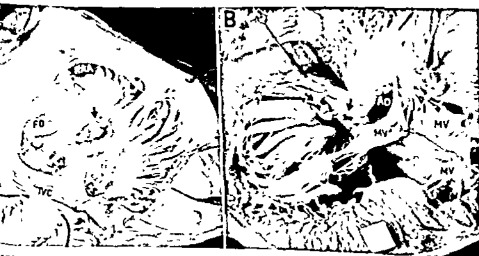
In the third case, the picture was dominated by great enlargement of the right atrium and considerable dilatation of the pulmonary artery and its left main branch, as well as greatly reduced blood volume in the lungs The left atrium was not enlarged

In the fourth case, the roentgenologic appearance was typical in every respect of tetralogy of Fallot.

Reduced vascularity of the lungs was thus the only feature present in every case. *If the right atrium is greatly enlarged in* such cases, the possibility of stenosis of the tricuspid valve must be considered When the atrium is emptied through associated



51b —Tricuspid stenosis, subvalvular pulmonary stenosis, single ventricle, and dextro-
oy, aged 14 (A.A. 380728). Right auricular appendage is large and causes a bulge in



651b — Same case as in Figure 651a.



Fig. 652.—Tricuspid stenosis and valvular pulmonary stenosis. Girl, aged 9 (E.H 430128), see Figure 657. *A*, at age 9; *B–D*, at age 11 Considerable enlargement of right atrium; its appendage shows long surface contiguity to anterior wall of the thorax (arrow in *D*). Caudad to it, there is a large space between the anterior wall of the heart and the thorax. Relative heart volume increased somewhat between examinations Reduction in vascularity of lungs

defects, no enlargement can be observed, and roentgenologic examination gives no direct reason for one to suspect the presence of tricuspid involvement.

CARDIAC CATHETERIZATION

In uncomplicated tricuspid stenosis, there is a pressure gradient between the right atrium and ventricle during ventricular diastole. Since congenital tricuspid stenosis is almost invariably combined with other anomalies, the pressure gradient does not usually appear. The pressure in the right atrium may be relieved by an atrial septal defect The flow through the tricuspid valve is therefore small, and if there is a ventricular septal defect, the right ventricle may be partly filled from the left ventricle Moreover, in severe tricuspid stenosis, it may be impossible to pass the catheter through the orifice into the ventricle. Under such conditions, stenosis cannot be distinguished from atresia. It is then necessary to complement the investigation with angiocardiography, not least in order to establish the nature of the coincident malformations

Cardiac catheterization was performed in our four cases (Table 24). In one the catheter did not pass through the tricuspid orifice At the time the investigation was made, our laboratory was not equipped with apparatus for angiocardiography The diagnosis was only made post mortem. In the other three cases, the investigation was more complete However, in only one case was it possible to advance the catheter into the pulmonary artery. When attempts were made to pass the infundibulum in the other two, bursts of ventricular extrasystoles occurred, and the catheter was therefore withdrawn It was possible by means of angiocardiography to diagnose valvular pulmonary stenosis in two cases and tetralogy of Fallot in one

The pressure curve from the right ventricle has a characteristic appearance (Figs 653, A, and 654, A) Although the ventricle works against increased resistance—in the form in one case of pulmonary stenosis and

TABLE 24.—Tricuspid Stenosis: Findings on Cardiac Catheterization in 4 Cases*

Case	O₂ Content, vol %									O₂ Cap., vol %	Pressure, mm Hg						
	SVC	IVC	RA	RV	PA	LA	PV	LV	Fem A		RA Mean	RV Syst	RV End-diast	PA Mean	LA Mean	LV Syst	LV End-diast
A A 380728	17.0	—	19.2	—	—	—	28.4	22.4	—	29.0	0	—	—	—	—	140	0
E H 430128	18.0	19.5	18.6	18.0	—	22.4	28.1	22.4	22.1	29.5	7	92	8	—	5	131	12
L E 461207	12.2	11.5	11.5	11.5	11.1	15.7	20.4	16.6	—	21.4	4	68	4	8	4	75	4
D B 460508	17.2	18.4	17.5	17.8	—	22.8	28.6	21.3	—	29.2	5	91	6	—	5	101	11

* For abbreviations see Table 1, p 119

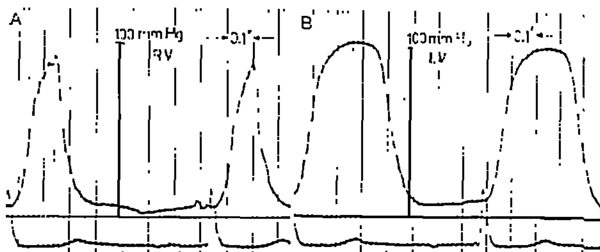


Fig. 653.—Tricuspid stenosis combined with tetralogy of Fallot. Girl, aged 8 (D.B. 46058 Pressure curve from the right ventricle (*A*) shows a shortened ejection phase, whereas that fro the left ventricle (*B*) shows this phase to be somewhat prolonged.

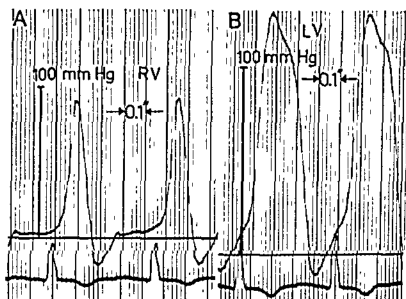


Fig. 654.—Tricuspid stenosis with valvular pulmonary stenosis. Girl, aged 11 (E.H. 430128) Pressure curves from the right (*A*) and the left (*B*) ventricle show changes similar to those in Figure 653.

in one of an over-riding aorta—the ejection phase is strikingly short, indicating small diastolic filling The large shunt from the right atrium to the left results in increased diastolic filling of the left ventricle, which has a considerably longer ejection phase (Figs 653, B and 654, B).

Decreased filling of the right ventricle and a fall in the stroke volume may also be caused by right ventricular failure It is nevertheless hardly probable that the right ventricle would become incompetent at this age, in the presence of such mild pul-

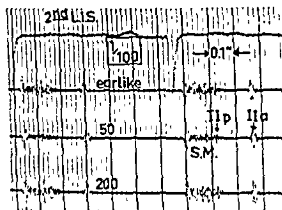


Fig 655.—Phonocardiogram in tricuspid stenosis ... 460508) Systolic murmur *(SM)* over the ... sound, which is the pulmonary componen... the short right ... Aortic compon... phase Boxed fi... the filters

monary stenosis and tetralogy of Fallot, respectively. The normal end-diastolic pressure in the right ventricle is a further argument against ventricular failure

The shortened ejection phase in the right ventricle is reflected on the phonocardiogram It is particularly apparent in case O.B 460508 (Fig. 655), in which the blood from the ventricle could escape into the over riding aorta The aortic component of the second sound is slightly late, but the most striking feature is that the pulmonary component occurs abnormally early and is extremely faint, inaudible to the ear The systolic murmur, caused by the blood flow through the pulmonary stenosis, is short but fills the whole interval between the first sound and the pulmonary component of the second sound. The duration of the murmur corresponds to the ejection phase of the right ventricle (Fig. 653).

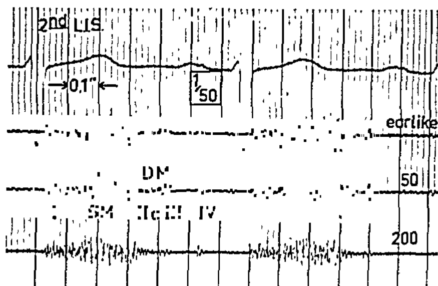
In cases E.H. 430128 and L.E. 461207, the systolic murmur was louder and lasted until the aortic component (Fig. 656). In pulmonary stenosis, the pulmonary component usually occurs considerably later than the aortic component, but in these cases the former could not be distinguished It probably occurred earlier and was masked by the murmur, which could not, therefore, be due to pulmonary stenosis alone. In case E.H. 430128 a sound was recorded 0.10 sec after the second sound. It could not be an opening snap of the tricuspid valve, since the summit of the *v* wave in the right atrium occurred before the second sound. It was probably a loud third sound due to dilatation of the left ventricle. Moreover, a faint protosystolic murmur was present

ANGIOCARDIOGRAPHY

Angiocardiographic examination was performed in *three cases* In one in which tricuspid stenosis was combined ...

ogy of Fallot, selective angiocardiography was used, in view only of the latter malformation. The contrast medium was injected into the right ventricle; consequently, the tricuspid stenosis was not visualized.

In two other cases (Fig. 657), the contrast medium was injected into the right atrium. Frontal and lateral projections were chosen, since, in view of the fact that the level of the systolic pressure was the same in both ventricles, it was considered essential to establish the position of the aortic root. It is possible that these projections were not ideal for study of the stenosed atrioventricular valve However, because of the anteromedial position of the anterior segment of the atrium, it is uncertain whether the orifice would have been visualized better in oblique projections. The two cases exhibited almost identical features. The right atrium emptied into the right ventricle by means of a narrow stream of contrast medium. The ventricle was small and its wall greatly thickened Typical valvular pulmonary stenosis and a distinctly dilated pulmonary artery were depicted An interatrial communication was present in one of them. The examination gave no grounds for suspecting the presence of a communication between the ventricles. In both cases the left atrium and ventricle and the aorta had a normal shape and size. Filling of a small right ventricle via the right atrium, when the left atrium and ventricle have not yet been visualized, affords strong indirect evidence of the existence of tricuspid stenosis as well. It is, of course, not improbable that similar features could be observed in primary hypoplasia of the right ventricle without stenosis of the tricuspid orifice (cf. pp. 736 and 737).



... pulmonary stenosis and patent ... SM) over the 2nd left interspace ... the 2nd sound (*IIa*). No pulmo- ... present, *as well as a 3rd heart* ... ably of atrial origin (*IV*) Boxed ... ndard frequencies of the filters

UNDERDEVELOPED RIGHT VENTRICLE WITHOUT TRICUSPID ATRESIA OR STENOSIS

We had one case of an underdeveloped right ventricle without tricuspid stenosis. There was a coincident large ventricular septal defect, both infundibular and valvular pulmonary stenosis, a patent foramen ovale and a patent ductus arteriosus. Complete diagnosis was made only at autopsy.

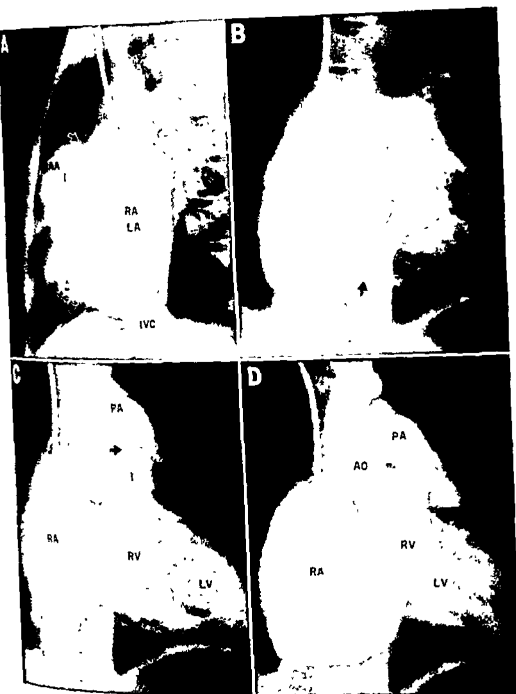


Fig 657 —Tricuspid stenosis and valvular pulmonary stenosis Girl, aged 9 (E.H 430128). Arrow in B points to a thin, membranous filling defect, it represents either the anterior medial border of right atrium or the atrioventricular border. [illegible] tricle small ar[illegible] atrium and d[illegible] Aorta is not [illegible] cava, LA and [illegible], left and right atria, LV and RV, left and right ventricles, PA, pulmonary artery, RAA, right auricular appendage

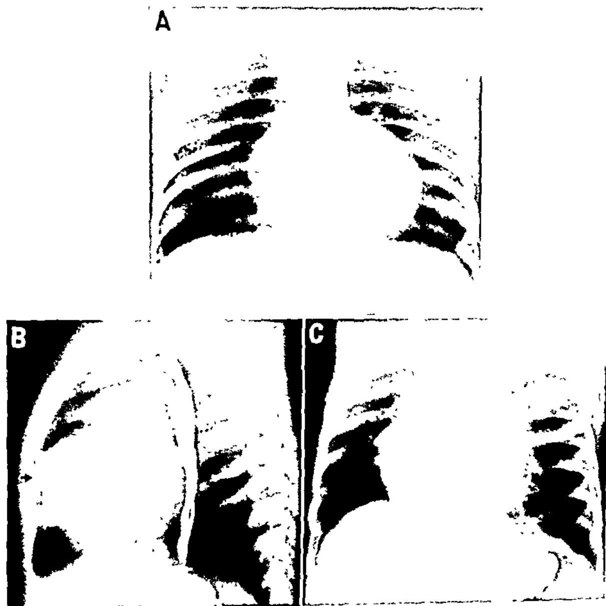


Fig. 658.—Underdeveloped right ventricle and patent foramen ovale. Girl, aged 1 yr. (K.E. 510426). Heart has same appearance as in tricuspid atresia, with considerable enlargement of right atrium and left ventricle Right auricular appendage is contiguous to anterior wall of the thorax (arrow in *B*) Greatly reduced vascularity of lungs.

Girl, aged 1 year (K.E. 510426).—A murmur was noted immediately after birth, but development was normal until 7 months of age, when she began to have attacks of cyanosis and loss of consciousness Successive exacerbation took place and she became permanently cyanotic On examination, she presented moderate cyanosis but neither dyspnea nor edema The cardiac findings were characterized by a very loud, long systolic murmur which lasted throughout the second sound, it had a maximum over the second left interspace No precordial bulge was present

Electrocardiography—This showed left ventricular hypertrophy, but also signs of right ventricular activity, although less than normally.

Roentgenologic examination. — The heart was moderately enlarged (Fig 658), the right atrium was greatly dilated and was contiguous to the anterior wall of the thorax. The ventricular portion had a characteristic shape; there was considerably increased curvature with a slight prominence in the superior lateral segment, indicating that it was formed chiefly by the left ventricle. The ventricle was greatly dilated and enlarged.

The vascularity of the lungs was sparse and

the pulmonary artery could not be identified The appearance was thus in good conformity with the assumption of an underdeveloped right ventricle

Cardiac catheterization (Fig 659).—The catheter was introduced through the saphenous vein. It could not be passed through the tricuspid orifice but could on the other hand, be advanced into the left atrium and ventricle No right to left interatrial shunt was found, but a right to left interventricular shunt was present The blood had presumably passed, during diastole, from the right atrium through the right ventricle to the left ventricle

Angiocardiographic examination.—This was performed by injection of the contrast medium into the greatly enlarged right atrium through a catheter with the tip directed toward the foramen ovale. The contrast medium passed into the left atrium, and the left ventricle was visualized immediately, whereas no right ventricle could be identified Only in retrospect was it observed that an extremely small right ventricle had been depicted in the first exposures in the series The tricuspid orifice was patent, but it could not be established whether or not stenosis was present No satisfactory filling of the pulmonary artery was obtained This would have required complementary angiocardiography with injection into the left ventricle, this examination was not made No patent ductus arteriosus was visible

Autopsy.—The patient died one month later Autopsy was performed at the Pediatric Clinic in Gothenburg, the autopsy report has kindly been placed at our disposal by Dr L-E. Carlgren. There were considerable hypertrophy and moderate dilatation of the left ventricle and right atrium. The tricuspid orifice was of about normal width and led to a small ventricle (about 1.5 × 1 cm), this, in turn, communicated with the left ventricle through a large defect in the muscular part of the ventricular septum. Circumscribed infundibular stenosis (3 mm in diameter) was found in the right ventricle, with dilatation of the infundibulum into a small, thin-walled third ventricle The pulmonary artery was of practically normal width, and the ductus arteriosus was patent

Hypoplasia of the right ventricle without tricuspid stenosis has been described by Cooley *et al* (159). In one case the right coronary artery was lacking, but in another "there was no obvious reason for the right ventricle hypoplasia."

In order to explain the findings on catheterization, it is necessary to assume that the small right ventricle functioned as a channel through which the blood flow passed, during diastole, from the right atrium to the left ventricle. There was, in fact, no right to left interatrial shunt. The passage

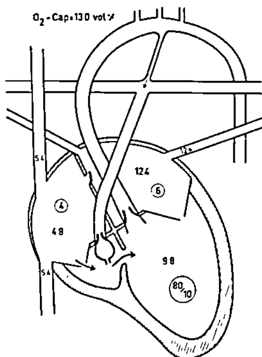


Fig. 659.—Underdeveloped right ventricle

others denote O_2 content in volumes %.

of the contrast medium from the right atrium to the left atrium during angiocardiography was due to the fact that the injection was made with the tip of the catheter directed toward the foramen ovale. No such sequence of events is found in uncomplicated atrial septal defect, but it may take place in cases in which the small right ventricle offers high resistance to the flow

This rare malformation is difficult to distinguish from tricuspid stenosis or atresia, a fact also stressed by Cooley *et al* (159)

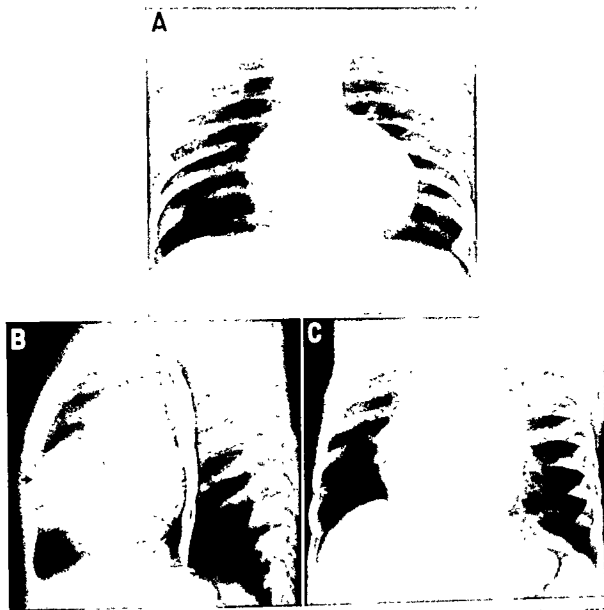


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The vascularity of the lungs was sparse an

TABLE 25 —MITRAL ATRESIA ANATOMY OF ASSOCIATED MALFORMATIONS, SYMPTOMS AND CLINICAL FINDINGS IN 7 CASES

...ase	Associated Malformations*	General Condition	Cyanosis	Liver	Cardiac Findings
520823 ...rl, ...mo)	*Angio* + *Cath.* + *Autopsy* Communication between atria via coronary sinus + single ventricle + patent ductus	Underdeveloped, easily fatigued; death at age 11 mo	Severe since birth	Large pulsating	Precordial bulge; acc. 2nd sound over pulm area; no murmur
511101 ...rl, ...mo.)	*Angio* + *Autopsy* Complete anomalous venous return to RA + rudimentary LA + large RV + rudimentary LV + small VSD + valv. pulm stenosis + origin of aorta and PA from RV	Spells of unconsciousness, easily fatigued; death at age 7 mo.	Severe since birth	Normal	Acc. pure 2nd sound over pulm. area; harsh systolic murmur, 4th L.I S
490814 ...oy, ...yr.)	*Angio* + *Cath* ASD + VSD + transpos of great vessels + large RV + small LV	Walks max 30–40 m on the level	Severe since birth	Slightly enlarged	Acc pure 2nd sound over pulm area; harsh systolic murmur over whole precordium
551221 ...oy, 1 yr ...mo.)	*Angio* + *Cath* Common atrium + single ventricle + transpos of great vessels + stenosis or atresia of left pulmon artery branch	Underdeveloped; dyspnea	Severe since birth	Normal	Murmur late in systole, 2nd R.I S
491124 ...rl, ...r)	*Angio* + *Cath* Dextrocardia + small ASD + transpos of great vessels + large RV with small outlet chamber (LV?) from which PA takes its origin	Normal development; walks max 200 m	Severe since birth	Normal	Precordial bulge, 2nd sound normally split, high-frequency diastolic murmur, 4th L.I S
460719 ...oy, ...yr)	*Angio* + *Cath* All pulmonary veins opening into RA + ASD + large VSD with over riding aorta + small LV	Retarded development, walks max 300 m; dyspnea	Slight	Normal	Precordial bulge; acc 2nd sound; grade 4 pansystolic murmur + low-frequency diastolic murmur over apex
551016 ...rl, ...mo)	*Autopsy* Common atrium + large RV + small LV + large VSD + patent ductus arteriosus + aplasia of inf vena cava (lower part drained by hemiazygous v) + 10 accessory spleens	Severe dyspnea, edema, death at age 3 mo	Slight	Marked enlargement	Grade 4 systolic murmur

...ath. = cardiac catheterization. Angio = angiocardiography

...n enlarged liver is as common a finding ...n tricuspid stenosis The second heart ...nd is usually accentuated, owing to ...nsposition of the aorta or pulmonary hy...tension The presence of a murmur is ... to an associated malformation, such as ...monary stenosis or ventricular septal ...ect. A murmur may be lacking. A dias...c murmur is sometimes heard, probably ... to the large flow through the tricuspid ...ice

ELECTROCARDIOGRAPHY

The ECG varies according to the nature of the associated malformations. All except one of our cases showed right ventricular hypertrophy ...

... small, rudimentary, non-functioning blind pouches In one case, S P. 551221, the ECG

25 Mitral Atresia

CONGENITAL MITRAL atresia is considerably less common than tricuspid atresia. From the functional standpoint they are similar, in that in both malformations there is a cor triloculare biatriatum, with complete arteriovenous mixing of the blood in the heart.

If a large atrial septal defect is present, the left atrium can be drained without difficulty into the right atrium. Edwards (216) has described the autopsy findings in five cases in which the communication between the atria consisted of a patent foramen ovale. Under such conditions, there is difficulty in emptying the left atrium. The pressure rises, the foramen ovale is dilated, and the valve is forced over to the right atrium, thus permitting a left to right shunt A ventricular septal defect allows the flow to pass from the right ventricle to the left. A single ventricle is sometimes present. The great vessels are often transposed.

As a rule, the pulmonary flow is decreased The high pressure in the left atrium results in pulmonary hypertension and increased resistance in the pulmonary vascular bed, thus causing a diminution in the pulmonary flow.

Mitral atresia is sometimes combined with atresia of the aortic valve, the left ventricle is then a nonfunctioning cavity (266). The systemic flow is maintained by the pulmonary artery through the ductus arteriosus. Patients with such malformations survive for at most a few days aft birth (650).

Our series contains seven cases of mitr atresia. The anatomic diagnosis and symp toms and signs are assembled in Table 25 The diagnosis was verified at autopsy i three cases. Transposition of the great ves sels was present in five cases. Two patient had complete anomalous venous return.

CLINICAL FEATURES

All but one of our patients were cyanoti from birth Even three patients without pul monary stenosis were severely cyanotic Since there was a marked impediment t emptying the left atrium, the pulmonary flow was decreased. In case E.L. 520823, the foramen ovale was sealed, and the only communication between the atria consisted of an opening between the left atrium and the coronary sinus. It was only 5 × 5 mm and was insufficient for effective drainage of the left atrium. All of the pulmonary veins opened normally on the left side. The patient was deeply cyanotic. On the other hand, cyanosis was only slight in one patient with complete anomalous pulmonary venous drainage and in another with a common atrium

Severe disability characterized all of our cases. Three patients died during the first year of life The oldest patient was aged 9 years, in this case all the pulmonary veins opened into the right atrium.

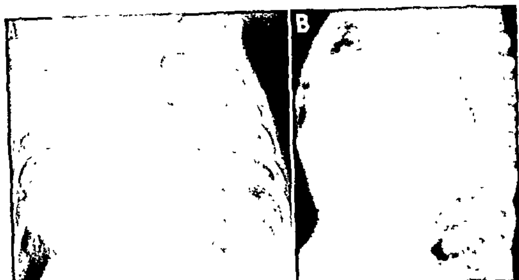


Fig 661.—Mitral atresia, drainage of left atrium through the coronary sinus, single ventricle, and patent ductus arteriosus Girl, aged 7 months (E L 520823), see Figure 666. Greatly en-
ventri-
to the

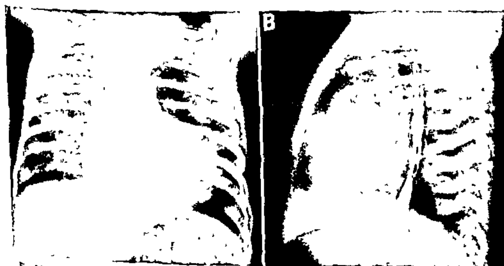


Fig 662.—Mitral atresia, complete abnormal venous return to right atrium rudimentary left ventricle, valvular pulmon...
sistent left superior ...

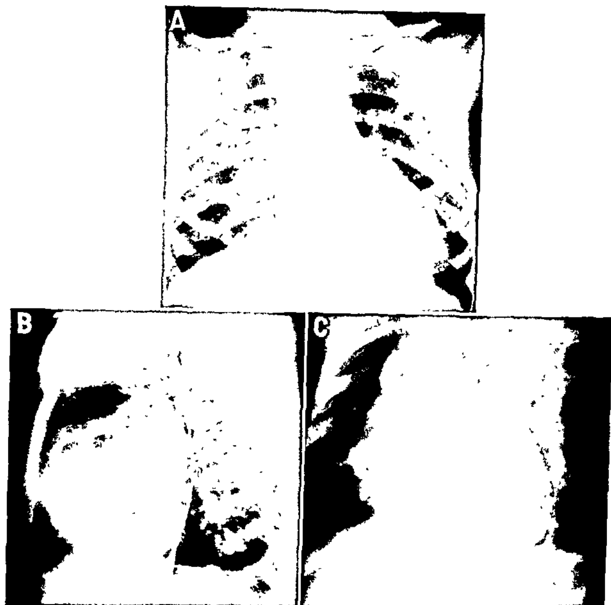


Fig. 660.—Mitral atresia, transposition of great vessels, and rudimentary left ventricle Boy, aged 3 (G A 490814), see Figure 665 Enlargement of right atrium and ventricle, shape of the latter is hypertrophic Narrow vascular pedicle, as in transposition of the great vessels; considerably increased vascularity of lungs, no enlargement of left atrium.

showed no definite ventricular hypertrophy, a single ventricle was present

ROENTGENOLOGIC EXAMINATION

The roentgenologic findings in this condition are unspecific and presumably seldom give rise to suspicions of mitral atresia. The roentgenologic appearance is highly variable, due to the multiplicity and divergent nature of the associated malformations. The size of the left atrium in particular is subject to great variations, depending on its draining conditions and the possible occurrence of anomalous venous return These variable features are illustrated by three of our cases (Figs. 660–662).

Boy, aged 3 years (G A. 490814, Fig 660) —The heart, which was moderately enlarged, had a strikingly narrow vascular pedicle with no distinct aortic arch segment The pulmonary artery was not visualized, but the lungs had abundant vascularization. The right atrium and ventricle were enlarged, but the left atrium was not dilated. The appearance was

artery These conditions also apply in complete anomalous venous return to the right atrium. The only difference is that, in the latter malformation, samples from the left atrium also have the same oxygen content as the aforementioned vessels, whereas in mitral atresia, saturated blood is found. In our cases, the catheter did not pass into the left atrium Moreover, the results of gas analyses are somewhat unreliable in infants with severe cyanosis The saturation of the blood may vary considerably from one moment to the next, and all the blood there is also a rise in pressure in the right atrium, with giant *a* waves fully comparable to those seen in tricuspid atresia (Fig. 663).

It is scarcely possible to diagnose mitral atresia with certainty by means of cardiac catheterization, even if the oxygen content and the pressure are determined in all the chambers of the heart and the great vessels. Complementary angiocardiographic examination is required, not the least reason for which is to establish the nature of the associated malformations that exist.

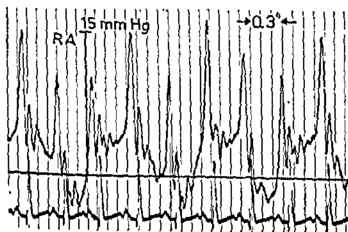


Fig 663.—Mitral atresia Girl, aged 9 months (E L 520823) Pressure recording from right ...um shows giant *a* waves

...ples cannot be taken simultaneously. ...t is sometimes possible to demonstrate ...sposition of the aorta by means of the ...tion of the catheter (*e g* , G A. 490814) ...case E L 520823, the catheter passed ...ugh the patent ductus into the descend- ...aorta The pressure was the same in the ...ta and in the pulmonary artery, and ...ce the oxygen content of the two vessels ...st also be the same, it could not be deter- ...ned whether or not a shunt was present ...murmur was audible At autopsy three ...nths later, only a narrow patent ductus ...s found (inner diameter at most 2 mm) The pressure in the right ventricle corre- ...nds to the systemic pressure The flow ...ough the right atrium is exceedingly ...ge and the resistance to filling of the ...pertrophied ventricle high. Consequently

ANGIOCARDIOGRAPHY

It should be possible at angiocardiography to establish the diagnosis by injection of the contrast medium into the left atrium. This could, however, be done in only one

[illegible] in a complementary examination with injection of contrast medium into the ventricle, this was shown to be single. Both the aorta and the pulmonary artery, which were transposed, were given off entirely from it.

In one case, intravenous injection showed only that the right atrium was enlarged and the great vessels transposed. Dilution of the

thus typical in every respect of transposition of the great vessels, and there was nothing to suggest the presence of mitral atresia as well. The diagnosis was based on correlation of the findings at cardiac catheterization and angiocardiography.

GIRL, AGED 7 MONTHS (E L. 520823, Fig. 661).—The heart volume was considerably increased. The outline was abnormal and the greatly enlarged atria showed an almost aneurysmal dilatation. The shape of the ventricular region was, on the contrary, uncharacteristic. The aorta was dilated, whereas the pulmonary artery was less prominent than normally. The vascularity of the lungs was increased. The dilatation of the left atrium was out of proportion to the increased blood flow in the pulmonary circulation, and it is reasonable to assume that this feature was due mainly to an impeded outflow. The considerable dilatation of the right atrium cannot be explained only on the grounds that the flow from both the systemic and the pulmonary circuits passed through it. The fact that emptying took place against higher resistance than normally must have been partly responsible (cf. the autopsy findings, p 749).

GIRL, AGED 7 MONTHS (S A 511101, Fig. 662).—In the moderately enlarged heart, the right ventricle was hypertrophied and exhibited a greatly curved anterior surface and an upturned apex The aorta was slightly dilated, whereas no part of the pulmonary artery could be visualized The vascularity of the lungs was sparse. The left atrium was not dilated and the autopsy findings (p. 746) showed that it was, in fact, rudimentary and, also, that all of the pulmonary veins opened into the right atrium

In the remaining four cases, the heart was also enlarged, a large left atrium could be established in three of them The vascularity of the lungs was increased in all four cases; in one of them, with atresia of the left pulmonary artery, the vascularity was sparse in the left lung and increased in the right.

CARDIAC CATHETERIZATION

Cardiac catheterization was performed in five cases, the results are recorded in Table 26. It is not possible to diagnose mitral atresia on the basis of these findings alone A large left to right interatrial shunt should be found, as well as the same oxygen content in samples from the right atrium, right ventricle, pulmonary artery, and a systemic

TABLE 26.—MITRAL ATRESIA: FINDINGS ON CARDIAC CATHETERIZATION IN 5 CASES*

Case	O₂ Content, vol %						O₂ Cap, vol %	Pressure, mm Hg									
	SVC	IVC	RA	RV	PA	Aorta		RA		RV		PA		Aorta			
						Asc.	Desc	Syst	Mean	Syst	End-diast	Syst	Diast	Asc Syst.	Asc Diast	Desc. Syst	Desc. Diast
E L 520823	7 5	7.5	8 2 9.7	11 3	10 7	—	10 5	16 0	13	4	65	6	62	44	—	—	67	46
G.A 490814	13 9	13 9	14 6	16 3	—	16 3	—	22 4	8	5	85	8	—	—	75	45	—	—
S P. 551221	9 7	9.7	10 4 12 9	10 2	—	—	—	18 0	9	6	77	9	—	—	—	—	—	—
M W. 491124	7 1	8 0	8 6	8 4	—	—	—	18 7	5	4	—	—	—	—	—	—	—	—
U A 460719	12 9	12 9	15 5	15 5	PV 18.1	—	Fem A 15 7	20.1	10	7	72	7	—	—	—	—	Fem A 88	45

*For abbreviations see Table 1, p 119

artery. These conditions also apply in complete anomalous venous return to the right atrium. The only difference is that, in the latter malformation, samples from the left atrium also have the same oxygen content as the aforementioned vessels, whereas in mitral atresia, saturated blood is found. In our cases, the catheter did not pass into the left atrium Moreover, the results of gas analyses are somewhat unreliable in infants with severe cyanosis The saturation of the blood may vary considerably from one moment to the next, and all the blood there is also a rise in pressure in the right atrium, with giant *a* waves fully comparable to those seen in tricuspid atresia (Fig. 663).

It is scarcely possible to diagnose mitral atresia with certainty by means of cardiac catheterization, even if the oxygen content and the pressure are determined in all the chambers of the heart and the great vessels. Complementary angiocardiographic examination is required, not the least reason for which is to establish the nature of the associated malformations that exist.

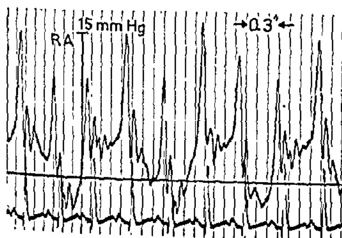


Fig 663 —Mitral atresia Girl, aged 9 months (E L 520823). Pressure recording from right atrium shows giant *a* waves

amples cannot be taken simultaneously

It is sometimes possible to demonstrate ransposition of the aorta by means of the osition of the catheter (e g, G A 490814) n case E L 520823, the catheter passed hrough the patent ductus into the descending aorta The pressure was the same in the orta and in the pulmonary artery, and ince the oxygen content of the two vessels must also be the same, it could not be determined whether or not a shunt was present No murmur was audible At autopsy three months later, only a narrow patent ductus was found (inner diameter at most 2 mm)

The pressure in the right ventricle corresponds to the systemic pressure. The flow hrough the right atrium is exceedingly arge and the resistance to filling of the ypertrophied ventricle high. O

ANGIOCARDIOGRAPHY

It should be possible at angiocardiography to establish the diagnosis by injection of the contrast medium into the left atrium. This could, however, be done in only one of our cases (Fig 664). It could be shown that the left atrium emptied exclusively into the right atrium...

...there, this was shown to be single. Both the aorta and the pulmonary artery, which were transposed, were given off entirely from it.

In *one* case, intravenous injection showed only that the right atrium was enlarged and the great vessels transposed. D...

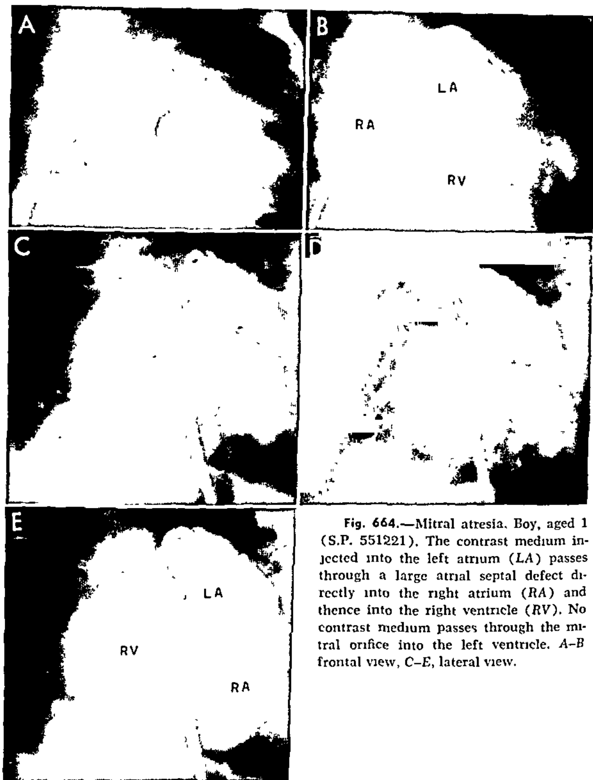


Fig. 664.—Mitral atresia. Boy, aged 1 (S.P. 551221). The contrast medium injected into the left atrium (*LA*) passes through a large atrial septal defect directly into the right atrium (*RA*) and thence into the right ventricle (*RV*). No contrast medium passes through the mitral orifice into the left ventricle. *A–B* frontal view, *C–E*, lateral view.

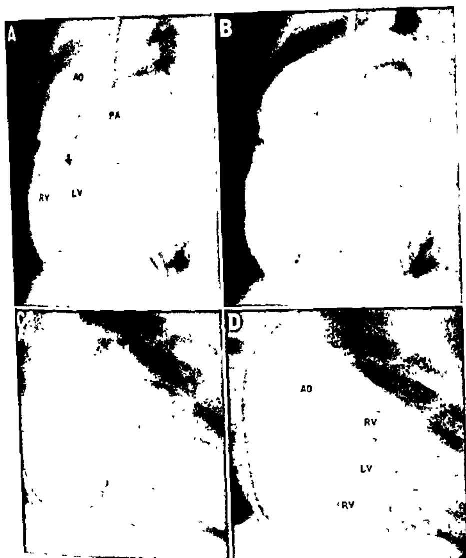


Fig 665 —Mitral atresia, transposition of great vessels, and rudimentary left ventricle Boy, aged 3 (G A 490814). Tip of the catheter is in the rudimentary left ventricle, which communicates through a 1 cm-wide septal defect with the large right ventricle (arrow in *A* points to defect) Pulmonary artery arises from the left ventricle, which lies behind the right During injection, the catheter tip recoils into the large right atrium, and the coronary sinus and cardiac veins become partly filled Owing to the right ventricular enlargement, the small cardiac vein (1 in F and G) is larger than the middle cardiac vein (*3* in *F* and *G*) *1*, great cardiac vein, 2, small cardiac vein, 3, middle cardiac vein, *Ad*, descending aorta, *AO*, aorta, *CS*, coronary sinus, *LV* and RV, left and right ventricles, *PA*, pulmonary artery, *RA*, right atrium (*continued*)

analysis. Selective angiocardiography was therefore performed, with injection of the contrast medium into the left ventricle; both the right ventricle and a wide ventricular septal defect were visualized at the same time (Fig. 665). The left ventricle was small and had the appearance of an outlet chamber. The pulmonary artery arose from it. No direct evidence of mitral atresia was afforded, since the left atrium was not visualized. The diagnosis was made on the basis of the underdevelopment of the left ventricle, despite its wide communication with the right ventricle, and the findings on cardiac catheterization

In a second case (Fig. 666), the first examination—using intravenous injection—showed considerable enlargement of the right atrium, whereas the details of the other parts of the heart were poorly visualized, owing to dilution of the contrast medium. When—in the second examination—the contrast medium was injected into the large ventricle, the aorta was clearly visualized and a patent ductus could also be identified Slight filling was obtained of an anteriorly placed small portion of the single ventricle. The pulmonary artery arose from this smaller cavity, which appeared on the angiocardiogram as an outlet chamber having a wide communication with the large ventricle. Since we were anxious to visualize the left atrium as well—which appeared from the ordinary roentgenologic examina-

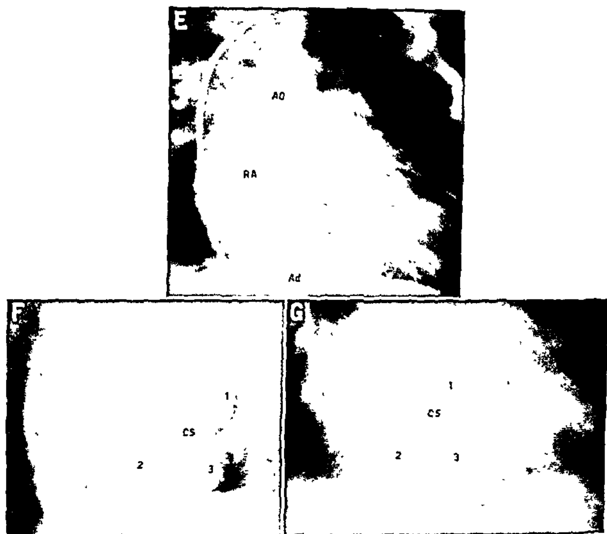


Fig. 665 (*cont*)

A

LV

RA

IVC

B

AO

LV

C

PDA

AO

Ad

D

AO

LV

Fig 666a —Mitral atresia [illegible] single ventricle and patent ductus arteriosus [illegible] B–D, left ventricle, E–H, [illegible] vein, communicates with an outlet chamber [illegible], which aorta runs in a curve d[illegible] on the right, is large (G[illegible] tion with the ventricle. [illegible] mitral orifice *Ad*, desc[illegible] aorta, *AO*, ascending aorta, *IVC*, inferior vena cava, *LA* and *RA*, left and right atria; *LV*, single ventricle, *PA*, pulmonary artery, *PDA*, patent ductus arteriosus, *RV*, right ventricle (outlet chamber), *VS*, ventricular septum (*continued*)

analysis. Selective angiocardiography was therefore performed, with injection of the contrast medium into the left ventricle, both the right ventricle and a wide ventricular septal defect were visualized at the same time (Fig. 665). The left ventricle was small and had the appearance of an outlet chamber The pulmonary artery arose from it. No direct evidence of mitral atresia was afforded, since the left atrium was not visualized. The diagnosis was made on the basis of the underdevelopment of the left ventricle, despite its wide communication with the right ventricle, and the findings on cardiac catheterization

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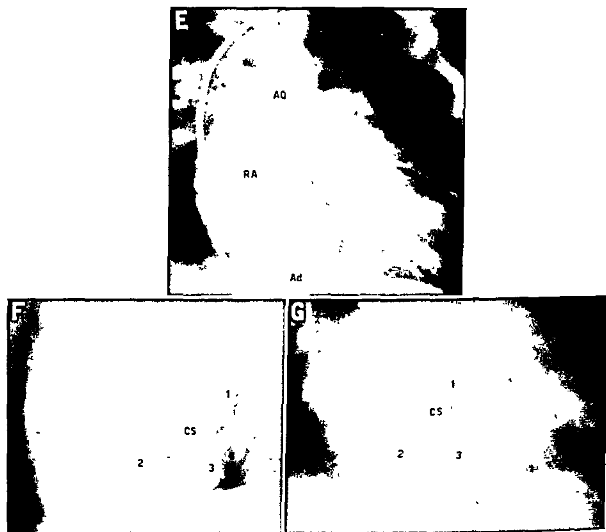


Fig. 665 (cont)







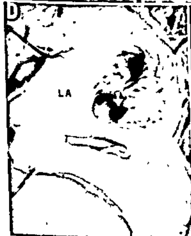




Fig. 666b —Same case as in Figure 666a Hollow (arrow in D) is seen at the ...
atresia Coronary ...
Foramen ...
pulmonar ...
ventricle ...
Malforme ...
cava, *LA* ...
auricular appendage, *RV*, right ventricle (outlet chamber), *TV*, tricuspid valve; *VVC*, valve of ... pulmonary artery; *RAA*, right
vena cava

tion to be grossly enlarged—an additional angiocardiographic examination was made. The contrast medium was injected into the outlet chamber, in order to obtain—via the return flow from the lungs—sufficient density in the left atrium. This was found to be

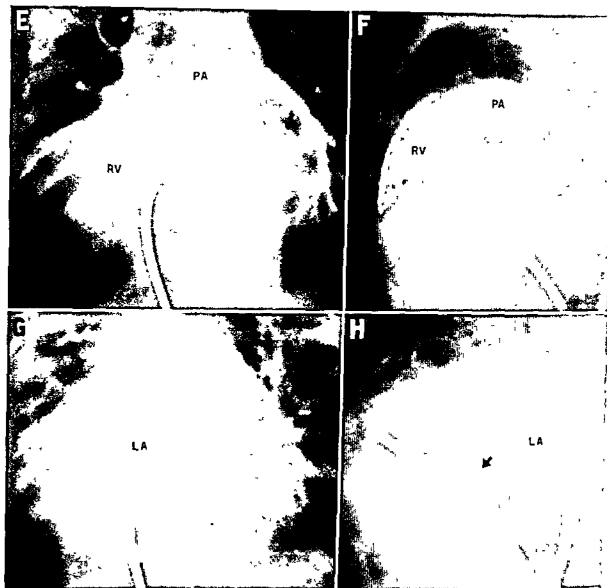


Fig. 666a (*cont*)

greatly enlarged, it showed inappreciable variations in volume and emptied slowly into the right atrium alone. The borderline between the left atrium and the large ventricle was sharply delimited, and a small bulge was visible in its medial segment. This possibly represented the site of the atresic mitral orifice (cf. the autopsy findings that are illustrated in Fig 666b)

In a third case, the contrast medium was injected intravenously. The right atrium, which was filled through a persistent left superior vena cava, was extremely large and partly occupied the site of the left atrium. The aorta was transposed and arose from a large right ventricle The pulmonary artery, which was inappreciably filled, arose to the right of the aorta and behind it The left atrium could not be identified. Autopsy disclosed that the left atrium was the size of a pea and that all of the pulmonary veins emptied into the right atrium. The mitral

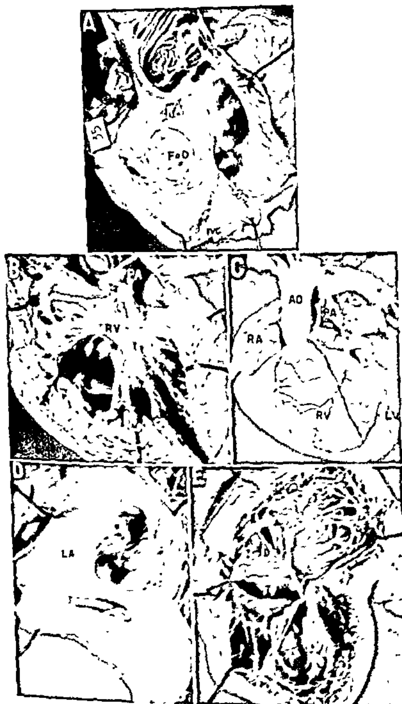


Fig 666b.—Same case as in Figure 666a. Hollow (arrow in *D*) is seen at the site of mitral atresia. Coronary sinus connects the left and right atria (probe in *A* and *D*) which are enlarged. Foramen ovale is closed. Tricuspid orifice opens into a single ventricle. The outflow tract to the pulmonary artery and adjacent part of the right ventricle is partly separated from the rest of the ventricle by abnormal columnar trabeculae. This smaller chamber lies forward and to the right. Malformed crista supraventricularis (*RV* in *B*). *AO*, aorta. *FoO*, fossa ovalis. *IVC*, inferior vena cava. *LA* and *RA*, left and right atria. *LV*, single ventricle. *PA*, pulmonary artery. *RAA*, right auricular appendage. *RV*, right ventricle (outlet chamber). *TV*, tricuspid valve. *VVC*, valve of vena cava.

tion to be grossly enlarged—an additional angiocardiographic examination was made. The contrast medium was injected into the outlet chamber, in order to obtain—via the return flow from the lungs—sufficient density in the left atrium. This was found to be greatly enlarged; it showed inappreciable variations in volume and emptied slowly into the right atrium alone. The borderline between the left atrium and the large ventricle was sharply delimited, and a small bulge was visible in its medial segment. This possibly represented the site of the atresic mitral orifice (cf. the autopsy findings that are illustrated in Fig. 666b)

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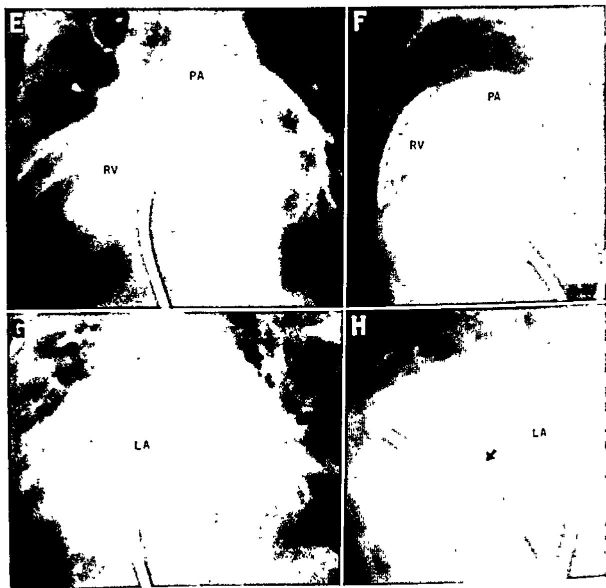


Fig. 666a (cont)

In one of the two remaining cases, selective angiocardiography showed that, during recirculation, the ventricles were opacified only through the right atrium, into which the left atrium emptied.

In the last case, the contrast medium was injected into the hemiazygos vein, which emptied into the coronary sinus (Fig. 667). The examination was so greatly hampered by the marked rotation of the heart that no decisive diagnostic data were obtained. The exact situation could be clarified only at autopsy.

To sum up, it can be stated that a diagnosis of mitral atresia can be established with most certainty on angiocardiography by injection of contrast medium into the left atrium when this chamber can be shown to empty only into the right atrium. If the pulmonary veins are transposed, the same phenomenon can be demonstrated after visualization of a small left atrium, filled from the opacified right atrium via an atrial septal defect. Lack of opacification of the left ventricle from the left atrium during recirculation is presumably of no decisive diagnostic value.



Fig. 667.—Mitral atresia and rotation anomaly Girl, aged 1 (M H. 551016). Contrast medium injected into hemiazygos vein, which probably drains the lower part of the body. It empties into the left superior vena cava The inferior vena cava cannot be identified

orifice was atresic. There was stenosis of the infundibulum of the hypoplastic left ventricle, which communicated with the right ventricle through a septal defect.

In one case, injection of contrast medium into the right atrium resulted in direct filling of a small left atrium as well This emptied solely into the right atrium. Complementary examination, with injection of contrast medium into the right ventricle, disclosed the presence of a ventricular septal defect, through which a small left ventricle was opacified. The aorta, which was narrow, showed an inappreciable degree of over-riding. On recirculation, good opacification of the right atrium occurred, without visualization of the left atrium It was therefore concluded that all the pulmonary veins were transposed

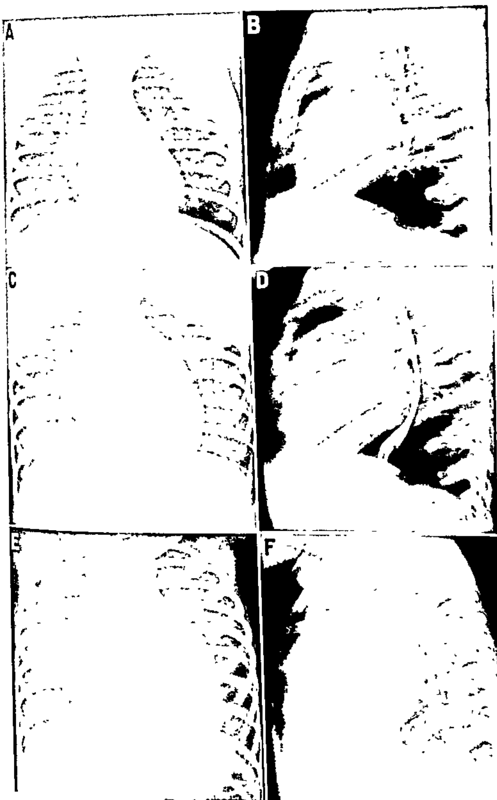


Fig 668a (legend on facing page)

26 Mitral Stenosis

ISOLATED CONGENITAL mitral stenosis is extremely rare. Ferencz *et al.* (251) found only 34 cases of congenital mitral stenosis published in the literature between 1846 and 1953 and added a description of nine new cases. Most of the cases reported were, however, combined with malformations of the aortic orifice or the aortic arch system, and only eight were entirely isolated. A few additional cases have been reported since then (86, 469, 502). Mitral stenosis is more common in association with atrial septal defect, but it is then considered to be of rheumatic origin. Occasional cases of the congenital type have nevertheless been described (581)

As a rule, the normal cusps are replaced by a funnel-shaped structure which projects into the often diminutive ventricle (216) The cusps are usually thickened and of a cartilage-like consistency. The chordae tendineae are deformed, short and thickened. Endocardial fibroelastosis of both the atrium and the ventricle has been demonstrated (82, 251, 469) Its pathogenesis is unknown. Some authors regard it as secondary to the stenosis (251), whereas others consider it to be a primary lesion (82, 469). If the endocardial thickening were present proximal to the stenosis, in the dilated atrium, it could conceivably be a secondary development, but this can scarcely apply to changes in the left ventricle in the absence of aortic stenosis. In some cases both the left ventricle and the aorta are hypoplastic, and fibroelastosis is not invariably present. Unequal division of the common atrioventricular canal has been suggested as the etiologic factor (82)

Congenital mitral stenosis usually gives rise to symptoms at an early age, generally at birth, and children affected rarely survive for many years. The symptoms are the same as those in acquired mitral stenosis Mild cases also exist, in which symptoms are lacking in childhood.

We have two cases in our series. One was of the severe type, the diagnosis was established clinically and verified at autopsy.

GIRL (G.W. 510415).—Birth weight was 3,250 Gm. She gained poorly in weight, at 13 months weighing 7,900 Gm A murmur was detected when she was 1 year old Both physical and mental development were retarded She could not stand alone until the age of 2 At this time she began to be dyspneic and also had a few cyanotic attacks She was never cyanotic in the interim Edema appeared a few months later After rapid exacerbation in the course of a week, with increasing edema and enlargement of the liver, she died at the age of 2 years and 5 months

at 1 yr.; *C* and *D*, at 1½
it ventricle and in total

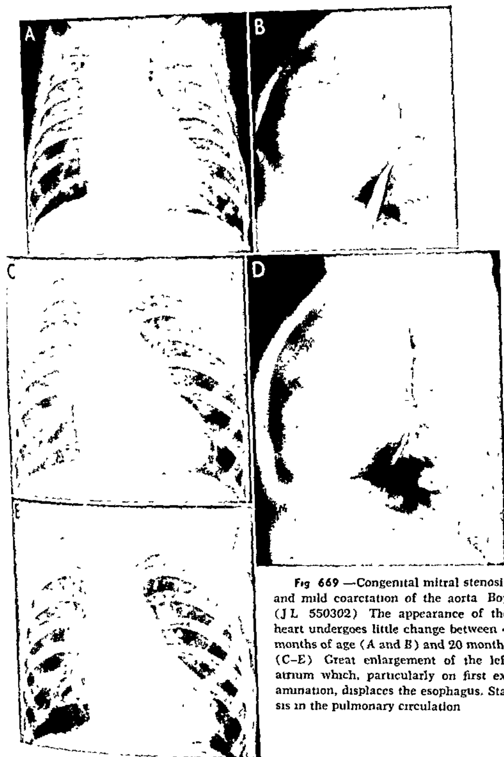


Fig 669 —Congenital mitral stenosis and mild coarctation of the aorta Boy (J L 550302) The appearance of the heart undergoes little change between 4 months of age (*A* and *B*) and 20 months (C–E) Great enlargement of the left atrium which, particularly on first examination, displaces the esophagus. Stasis in the pulmonary circulation

The *cardiac findings* were characterized by an accentuated first sound and a loud, long diastolic murmur over the apex.

The *electrocardiogram* showed notched P waves in leads I and II, but no right ventricular hypertrophy was recorded from the chest leads.

The first *roentgenologic examination* was made when the patient was 1 year old (Fig. 668a). The heart, which on repeated examinations during the second year of life was found to increase successively in size, had a typical outline. It was characterized by dilatation of the left atrium, a prominent auricular appendage, enlargement of the right ventricle and atrium, and dilatation of the central pulmonary vessels and the superior vena cava. The width of the aorta and pulmonary artery could not be estimated, since these vessels were partially overlapped by the thymus.

Cardiac catheterization was performed at 2 years of age, the results are seen in Table 27. A complete A-V block developed in the course o[f] catheterization, which was therefore stopped. (The ECG returned to its former state on th[e] following day.) Consequently, we obtained n[o] samples from the pulmonary artery, and th[e] PCV pressure could not be recorded. The arterial oxygen saturation was slightly decreased. Thus, catheterization gave little information of value for the diagnosis. The physical findings and roentgenologic appearance were nevertheless so typical that a diagnosis of mitral stenosis was regarded as unquestionable.



Fig. 668b.—Same case as in Figure 668a. Cusps of the mitral valve form a thick, irregular ring (*MV*) with a small central orifice (lower arrows). A biopsy specimen taken from the cusps

TABLE 27.—CONGENITAL MITRAL STENOSIS. FINDINGS ON CARDIAC CATHETERIZATION IN 2 CASES*

Case	O₂ Content, vol %						O₂ Cap, vol %	Pressure, mm Hg						
								RA	RV		PA		PCV	
	SVC	IVC	RA	RV	PA	Fem A		Mean	Syst	End-diast	Syst	Diast	a wave	Mean
G.W. 510415	8.4	10.7	8.7	9.0	—	15.8	19.5	4	50	3	61†	45†	—	—
J.L. 550302	10.8	12.1	10.1	10.2	10.5	14.2	14.5	2	30	4	29	11	15	9

*For abbreviations see Table 1, p. 119

†Complete A-V block appeared on pressure recording in PA. Catheterization was terminated, and PCV pressure was not recorded.

Angiocardiography was not performed, on account of the A-V block.

The diagnosis was confirmed at *autopsy* (Fig. 668b), which showed the cusps of the

what small but not hypoplastic. A number of

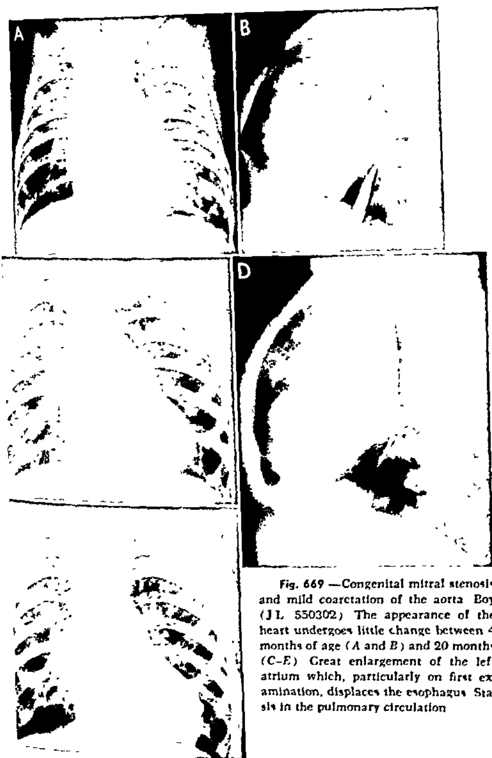


Fig. 669 —Congenital mitral stenosis and mild coarctation of the aorta Boy (J L 550302) The appearance of the heart undergoes little change between 4 months of age (*A* and *B*) and 20 months (*C–E*) Great enlargement of the left atrium which, particularly on first examination, displaces the esophagus Stasis in the pulmonary circulation

yellowish-white, thickened strands, probably endocardial fibroelastosis, were present in the *endocardium*. The right ventricle and atrium were dilated, and there was pronounced hypertrophy of the trabeculae. The atrial and ventricular septa were intact. The ductus arteriosus was obliterated.

The other patient had slight stenosis, associated with very mild coarctation of the aorta. The diagnosis was based on the physical findings and roentgenologic examinations.

Boy, aged 1 year 8 months (J.L 550302). —Development was normal and he was asymptomatic. A murmur was detected when he was *newborn.*

The *cardiac findings* were characterized by an apical presystolic murmur and a short, diamond-shaped murmur in late systole The coarctation, visualized on angiocardiography, was very mild. The femoral pulse was dis-

sometimes seen in coarctation of the aorta with a direct communication between the upper and lower segments of the vessel

The *electrocardiogram* showed an incomplete right bundle-branch block, but neither atrial nor ventricular hypertrophy

Roentgenologic examinations were made *repeatedly between the age of 4 months and* 2½ years. The picture was invariably the *same* It was characterized by distinct enlargement of the heart, entirely attributable to dilatation of the left atrium, a prominent auricular appendage, and suggested abnormal vascular markings in the lungs, with indis hili, as in venous stasis (Fig. 669). The vessels had a normal appearance.

Cardiac catheterization showed normal monary artery pressure. The mean PCV sure was normal, but the *a* wave was al mally high (Fig. 670).

Angiocardiography was performed by i tion of contrast medium into the pulmo artery. It is seen from Figure 671 that the atrium was greatly enlarged and that its varied considerably during different phase the cardiac cycle The left ventricle wa *ordinary size, and there was no definitely* normal increase in the thickness of its v There was suggested dilatation of the asc ing aorta. Coarctation, producing inapp *able narrowing of the lumen, was visible d* to the origin of the subclavian artery from aortic arch. Since the coarctation caused

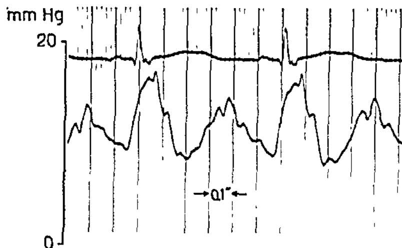


Fig. 670.—Congenital mitral stenosis Boy, aged 8 months (J L 550302). Pressure recor *in PCV position The mean pressure is normal, but the a waves are abnormally high*

marked hemodynamic changes, the consic able enlargement of the left atrium can be plained only by an impediment in the at ventricular plane *It was difficult to ju* whether fibroelastosis of the left ventricle v a contributory factor, but it seemed unlik in view of lack of thickening of the ventricu *wall and the distinct variations in size of t* ventricle during the cardiac cycle

Hilbish and Cooley (334a) *attach* great importance to stagnation of the co trast medium in the enlarged left atriu They *stated this phenomenon* to be s nificant of mitral stenosis, even in cas with coarctation of the aorta and hyperte sion and probable signs of left ventricul failure In our experience, however, it

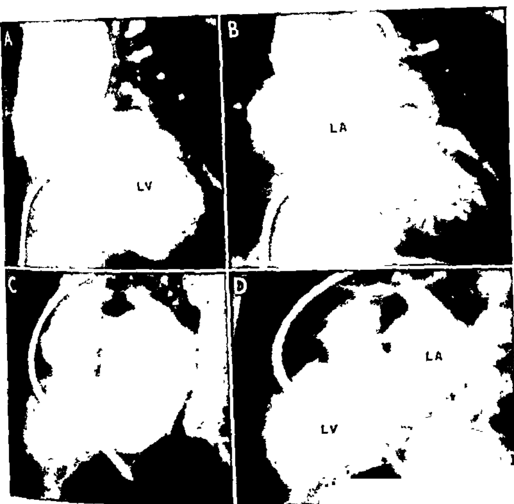


Fig. 671.—Congenital mitral stenosis and mild coarctation of the aorta (see Fig. 669). Boy, aged 1 (J.L. 550302). Contrast medium injected into pulmonary artery. The greatly enlarged left atrium (LA) empties very incompletely in atrial systole (*A* and *D*). Ordinary appearance of left ventricle (*LV*). The degree of mitral stenosis cannot be evaluated.

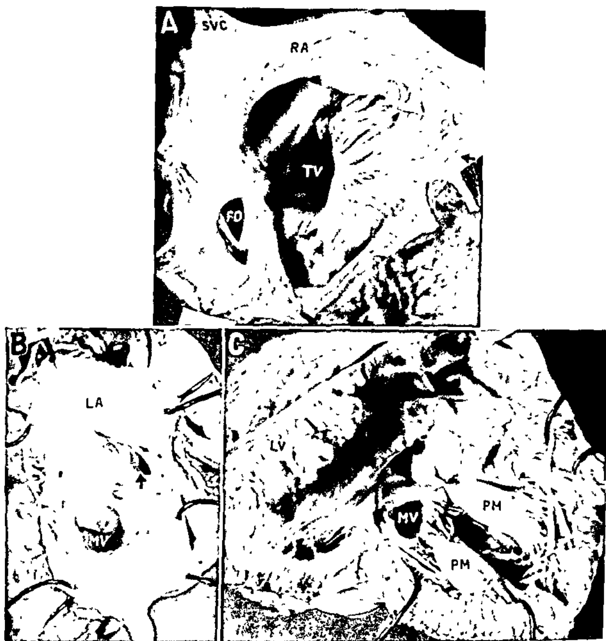


Fig. 672.—Mitral stenosis and patent foramen ovale Boy, aged 8 months (T G. 530420) Cusps of the mitral valve are fused into a funnel-shaped tube with an orifice about 7 mm in diameter (*MV* in *B* and *C*) Enlargement of the right atrium but not of the left. The valve does not entirely cover the foramen ovale (arrow in *B*). Chiari's net is seen in the right atrium (arrow in *A*) *FO*, foramen ovale, *LA* and *RA*, left and right atria, *LV*, left ventricle; *MV*, mitral valve, *PM*, papillary muscle, *TV*, tricuspid valve, *SVC*, superior vena cava

ustified to diagnose mitral stenosis in cases only on the basis of the afore-oned phenomenon.

ure 672 illustrates an autopsy speci-which does not belong to our series, ch the cusps are fused into an annu-embrane which greatly constricts the

view of surgical intervention, it is ial to determine whether the malfor-n is associated with hypoplasia of the entricle In this event, valvulotomy is indicated Definite estimation of the f the left ventricle can presumably be only by means of angiocardiography. ave had no experience of this examination in congenital mitral stenosis, but no difficulties have been encountered in visualizing the left ventricle in adults with acquired mitral stenosis. The contrast medium has then been injected either into the pulmonary artery or directly into the left atrium by the technique of percutaneous puncture (66).

Valvulotomy can be performed even in the presence of fibroclastosis. Two of the patients described by Bower *et al.* (82) were operated on, and one survived. There were also a wide patent ductus and a reversed shunt, and the effect of operation was therefore less evident. The patent ductus was not ligated

27 Mitral Incompetence

"DUPLICATION OF the mitral orifice" is a malformation consisting of an extra opening in one of its cusps. This might give rise to incompetence of the mitral valve, but it is generally regarded as being of no clinical importance (409, 543). Clinically manifest, congenital mitral incompetence is an exceedingly rare condition. Prior (543) has published an account of one patient who died at 26 years of age. Heart disease had *been detected only six years earlier, but the* autopsy findings were considered to be indicative of a congenital malformation. Prior wrote· "The free margin of the posterior leaflet was rolled, and there were no chordae tendineae from this leaflet to the papillary muscles." The foramen ovale was patent and only partly covered by the valve Edwards (218) described a case with a cleft mitral valve, in which heart failure and death occurred at 8 years of age Helmholz *et al.* (329) reported six cases with corrected transposition of the great vessels, combined with severe malformation of the left atrioventricular valve, which was incompetent.

Our series numbers three cases of mitral incompetence The heart disease was discovered so early—before 6 months of age—that it must be regarded as a congenital malformation In one of the patients (M N 450424) there was also corrected transposition of the great vessels.*

*In the previous edition of our book, this was described as a rotation anomaly.

CLINICAL FEATURES

The main symptoms and clinical finding are given in Table 28. Case M.N. 45042 was mild, but the two other patients ha severe signs and symptoms.

Cardiac Findings

Cardiac findings consisted of a left ven tricular thrust, a long, loud systolic murmu over the apex, a loud third sound, but r opening snap, and no accentuation of th first sound (Fig. 673) Case B.G. 38120 also had a faint mid-diastolic murmur ov the apex. The second sound over the pu monary area was accentuated in the tw severe cases, in which pulmonary hype tension was present. Right ventricular h pertrophy was so severe that it had pr duced a precordial bulge Cardiac failur and enlargement of the liver had alread developed in these cases, but peripher *edema was lacking*

Electrocardiography

The findings on patient M.N. 450424 ar discussed in connection with the electr cardiographic features in corrected tran position (see p 808) The other tw patients showed signs of pronounced le ventricular hypertrophy, as well as P m trale Although both had pulmonary hype tension, only one of them had signs of rigl ventricular hypertrophy as well (Fig. 674

ROENTGENOLOGIC EXAMINATION

Two of the cases presented a roentgenologic appearance which was, in all essentials, the same as that in advanced, acquired mitral incompetence The left atrium (Figs. 675 and 676) was aneurysmally dilated and the appendage was prominent, the right atrium and ventricle as well as the left ventricle were greatly enlarged The main trunk of the pulmonary artery and its central branches were dilated, whereas the peripheral vessels of the lungs were narrow, indicating increased resistance in the pulmonary circulation The aorta was markedly narrow, as an expression of a small output. No intracardiac calcifications were observed. The heart volume was greatly increased

In the third case (Fig. 677), there was a corrected transposition which made the interpretation more difficult. A comparison with the angiocardiogram showed that the greater part of the anatomical left ventricle lay to the right of the right ventricle and that the aorta, which was narrow, ascended on the left side. The pulmonary artery, which ran medially in the mediastinum, was not visualized on conventional roentgenologic examination The central pulmonary vessels were slightly dilated There was great dilatation of the left atrium, but its appendage was not prominent

The characteristic appearance in the first two cases—enormous dilatation of the left atrium, enlargement of the left ventricle, dilatation of the central lung veins, and peripheral changes in the pulmonary vessels indicative of increased resistance in this circuit—was highly suggestive of extremely severe mitral incompetence

The appearance was less specific in the

TABLE 28.—CONGENITAL MITRAL INCOMPETENCE: SYMPTOMS AND CLINICAL FINDINGS IN 3 CASES

Case	Age at			General Condition	Cyanosis	Liver Enlargement, Edema	Cardiac Findings
	Diagnosis of Heart Disease	Onset of Symptoms	Examination				
M.N 450424 (girl)	4 mo	5 yr	8 yr.	Physical devel. normal, able to run, on greater exertion, more dyspneic and easily tired than normal children	0	0	Slight precordial bulge, lat. displacement of apex beat, over apex, syst. thrill, long, loud syst murmur, loud 3rd sound
B.E. 461013 (girl)	6 mo	1–2 mo	6 yr	Poor weight gain in 1st year, never able to run more than short distance, worse in recent months, managing only 1 flight of stairs, dyspnea; normal height but underweight	0	Slightly enlarged liver, no edema	Marked precordial bulge, apex beat broad, displaced laterally; wide neck veins, over apex, long, loud syst murmur & 3rd sound, loud diast murmur over 3rd L I S, acc 2nd pulm. sound
B.C 381206 (girl)	3 mo	10 yr	13 yr.	Successive deterioration in past 3 yr, grade III disability, dyspnea, height normal but underweight	Slight, lips	Slightly enlarged liver, no edema	Marked precordial bulge, apex beat broad, displaced laterally, loud syst & faint diast. murmur over whole precordium, acc 2nd pulm. sound

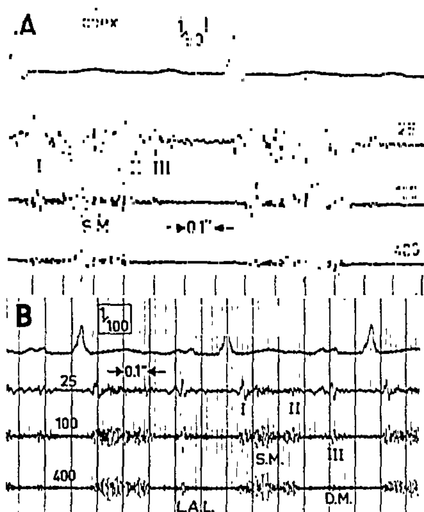
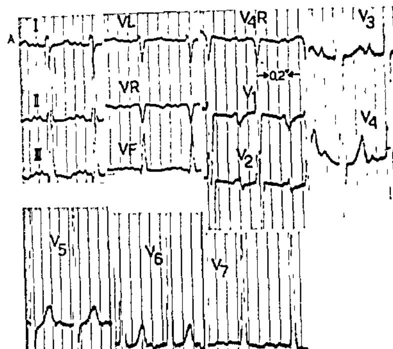


Fig. 673.—Phonocardiograms in two cases of mitral incompetence. *A*, girl, aged 8 (M.N 450424) A loud systolic murmur (*SM*) and broad, fairly high-pitched 3rd sound (*III*) are recorded over the apex *B*, girl, aged 6 (B E. 461013). A loud systolic murmur (*SM*), faint 2nd sound (*II*), broad 3rd sound (*III*) and possibly a mid-diastolic murmur (*DM*) are recorded over the left axillary line (*LAL*) at the level of the mammillae Boxed figures denote frequency of amplification, other figures denote standard frequencies of the filters.



674.—*A*, mitral incompetence. Girl, aged 6 (B E 461013), with moderately raised pres- the pulmonary artery (*continued*).

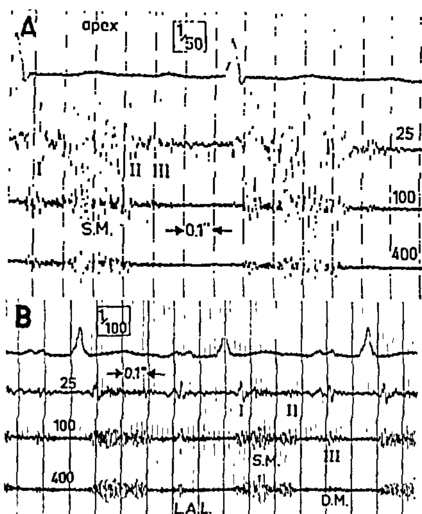


Fig. 673.—Phonocardiograms in two cases of mitral incompetence. *A*, girl, aged 8 (M.N. 450424). A loud systolic murmur *(SM)* and broad, fairly high-pitched 3rd sound *(III)* are recorded over the apex *B*, girl, aged 6 (B E 461013) A loud systolic murmur *(SM)*, faint 2nd sound *(II)*, broad 3rd sound *(III)* and possibly a mid-diastolic murmur *(DM)* are recorded over the left axillary line *(LAL)* at the level of the mammillae Boxed figures denote frequency of amplification, other figures denote standard frequencies of the filters

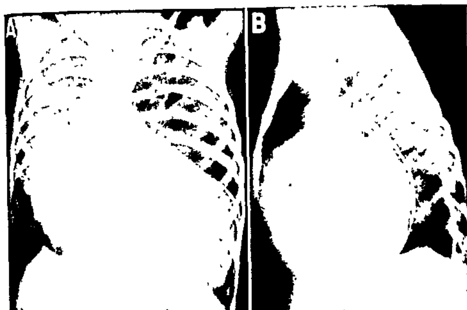


Fig 675 —Mitral incompetence. Girl, aged 6 (B E 461013) Heart volume is more than four times normal Left atrium is enormously dilated, forming the greater part of the right border. Considerable enlargement of the other chambers of the heart, extremely narrow aorta, great dilatation of arteries and veins centrally in lungs, peripherally, vessels are fairly narrow, middle lobe emphysematous

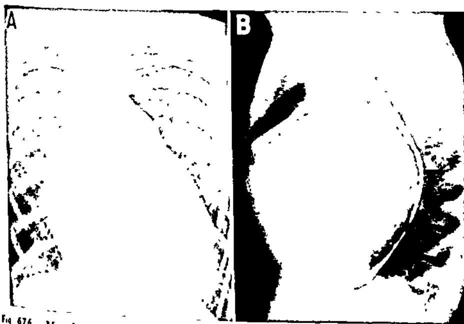


Fig 676 —Mitral incompetence Girl, aged 13 (B G 381206) Considerable increase in heart volume Great enlargement of all chambers of the heart, greatest of the left atrium, which forms part of the right border Marked dilatation of vessels centrally in the lungs, reduction peripherally, narrow aorta

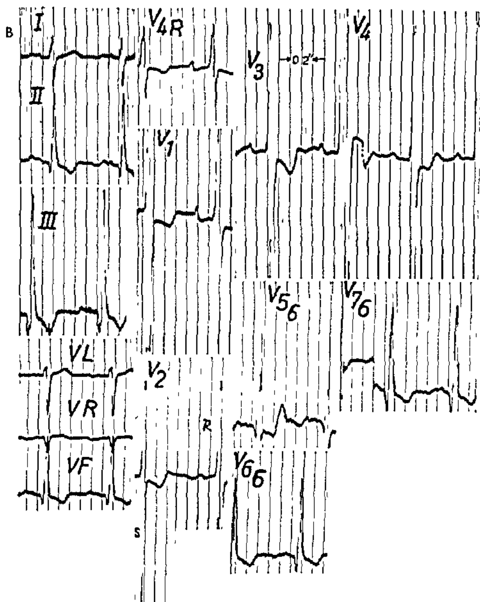


Fig. 674 (*cont*) —*B*, mitral incompetence Girl, aged 13 (B G 381206), with severe pulm nary hypertension

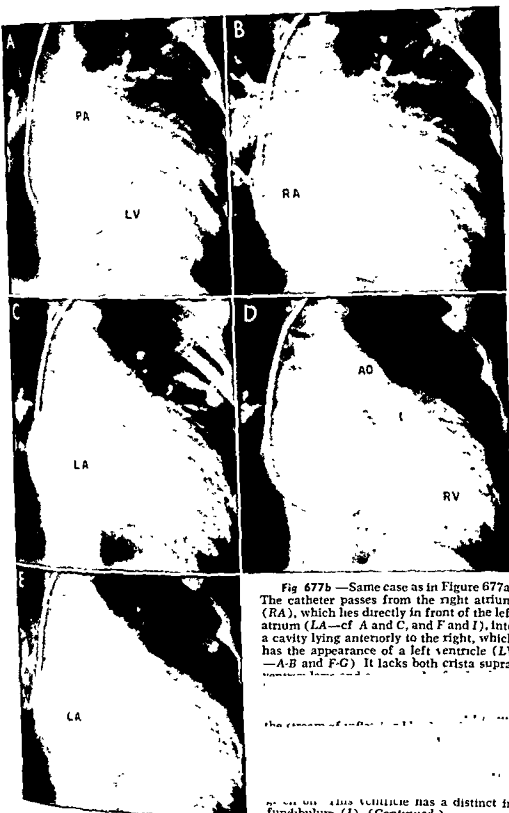
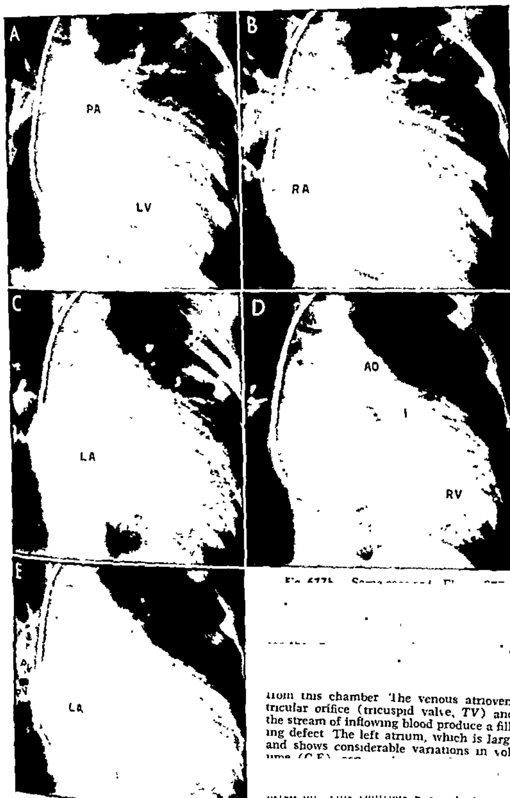


Fig 677b —Same case as in Figure 677a. The catheter passes from the right atrium (*RA*), which lies directly in front of the left atrium (*LA*—cf *A* and *C*, and *F* and *I*), into a cavity lying anteriorly to the right, which has the appearance of a left ventricle (*LV* —*A-B* and *F-G*) It lacks both crista supra-

This ventricle has a distinct infundibulum (*I*) (Continued.)



Fig. 677 ...
8 (M N. ...
aorta lies ...
cannot be identified The left cardiac outline is formed chiefly by the right ventricle. Slight [illegible]
monary stasis.



from this chamber The venous atrioventricular orifice (tricuspid valve, *TV*) and the stream of inflowing blood produce a filling defect The left atrium, which is large and shows considerable variations in vol-

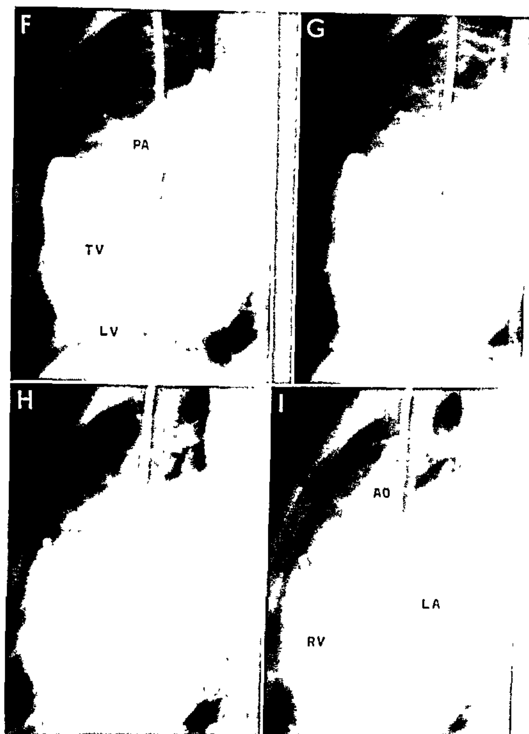


Fig. 677b (*cont*)

remaining case. By electrokymographic recording of the pulsations in the left atrium it was possible to make a tentative roentgenologic diagnosis of mitral incompetence

ELECTROKYMOGRAPHY

Seemingly typical findings in the electrokymogram of the left auricular appendage

ment to emptying, as usually recorded in mitral stenosis, could therefore be ruled out with a high degree of probability.

CARDIAC CATHETERIZATION

The findings obtained on cardiac catheterization are shown in Table 29.

In the mildest case (M.N. 450424), there

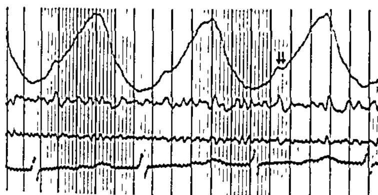


Fig 678 —Electrokymogram of left auricular appendage in congenital mitral incompetence Girl, aged 8 (M N 450424) The descent (between arrows) coinciding with the first part of the ventricular ejection phase is short · 0 02–0 03 sec. Continuous rise during the remainder of systole Normal downward deflection in early diastole

TABLE 29 —CONGENITAL MITRAL INCOMPETENCE FINDINGS AT CARDIAC CATHETERIZATION IN 3 CASES

Case	O₂ Content, vol %							O₂ Cap	Pressure, mm Hg					
	SVC	IVC	RA	RV	PA	PCV	FA		RA	RV	PA	PCV a	PCV v	PCV mean
M.N 450424	9 8	10 1	11 1	11.3	11 5	14 1	14 1 = 91%	15 5	5	32/5	27/14	15	20	15
B E. 461013	8 0	7 8	7 1	7 1	7 0	—	11 7 = 82%	14 3	1	65/5	57/5	—	—	18*
B G 381206	11.7	—	11 8	12 3	11 6	—	16 0 = 89%	18 0	5	85/5	70/30	—	—	20*

*Damped.

were made in the one case investigated They consisted of a shortening of the descent marking the movement of the atrioventricular plane toward the apex during the ejection phase of the ventricle, and a continuous rise during and beyond ventricular systole These features, as well as a normal downward deflection in early diastole, argued in favor of mitral incompetence (Fig. 678) On the other hand, no increased presystolic activity was present An impedi-

was normal pulmonary artery pressure but raised PCV pressure. The curve was not damped and the v waves were high (Fig 679) In the other two cases, the pulmonary artery and PCV pressures were both raised, but the curve of the latter was damped, as is usual when the resistance in the pulmonary vascular bed is high As a result of this, no tall v waves occur

A small admixture of arterial blood in the right atrium was found in case M.N

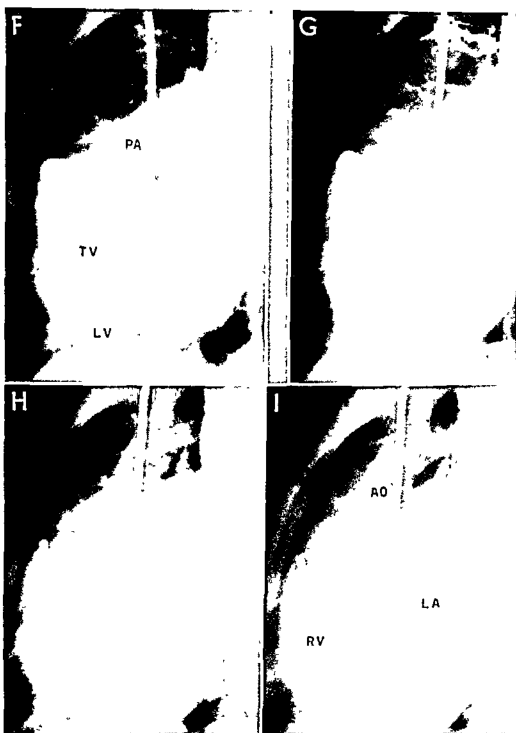


Fig. 677b (*cont*)

these patients (B E 461013) died in her home one year after examination; it was far from Stockholm and there was no opportunity of performing an autopsy. The other (B G. 381206) died two years after examination, autopsy was carried out at the County Hospital of Jönköping. The anterior cusp of the mitral valve was found to be lacking and the posterior cusp was rigid Both the left and the right ventricle were hypertrophied, and the left atrium was greatly dilated. The pathogenesis of this valvular defect may possibly be analogous to that in Ebstein's disease. In the latter condition, the malformation is considered to arise during the early embryonic stage, owing to incomplete regression and retraction of the connective tissue and musculature of the valvular primordium (35). In our case, in which the anterior cusp of the mitral valve was absent, this process may have failed entirely to take place.

450424, which would indicate an atrial septal defect or anomalous venous return to the right atrium. The shunt was small, but was regarded as significant; this view was substantiated by the findings on angiocardiography. The clinical picture was not, however, affected by this small shunt, the mitral incompetence being the dominating feature. There was probably a patent foramen ovale which, owing to the dilatation of the left atrium, was so greatly widened that the valve did not cover the opening and thus permitted a left to right shunt A case of this kind was described by Prior (543)

In all the cases the arterial oxygen saturation was slightly decreased, owing to deficient oxygenation of the lungs The arteriovenous oxygen difference was not high

ANGIOCARDIOGRAPHY

Angiocardiographic examination was done in one case (M.N. 450424), with the medium injected into the left ventricle (Fig. 677b) The pulmonary artery took off from this ventricle.

The left atrium was greatly dilated and the right ventricle, which was enlarged, became filled slowly. Slight opacification of the right atrium occurred simultaneously. No definite conclusions regarding the exact nature of the mitral valve lesion could be drawn, since optimal visualization of the orifice was not obtained with the projections used. Slow emptying of the left atrium, as in this case, is usually found in mitral stenosis, but the extent to which the modified emptying conditions in the presence of dominating mitral incompetence can be demonstrated by angiocardiography has not been determined The enlargement of the right ventricle is nevertheless an argument in favor of mitral incompetence The corrected transposition provides an explanation of the abnormal relative positions of the chambers of the heart.

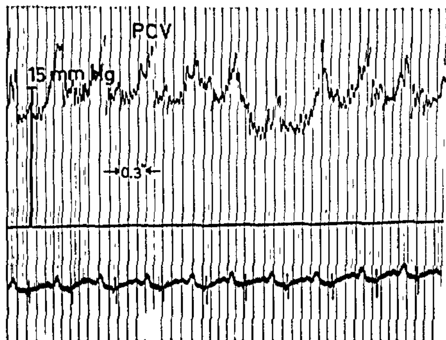
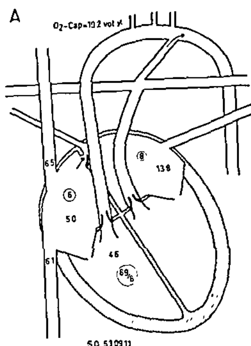
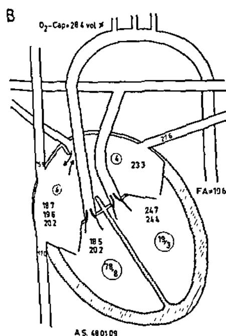


Fig. 679.—PCV pressure curve in mitral incompetence Girl, aged 8 (M N 450424) Normal pressure in right pulmonary artery and a small shunt from left to right atrium PCV pressure raised and *v* waves high

The two severe cases presented a characteristic picture The mitral incompetence was undoubtedly congenital Symptoms of heart failure appeared in early childhood, and the heart was enormously enlarged Consequently, it must have been a question, not of duplication of the mitral valve, but of severe malformation of its cusps One of





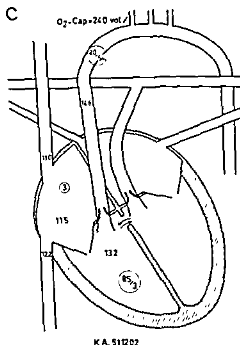


Fig. 680.—Examples of the hemodynamics in complete transposition of the great vessels. *A*, with patent foramen ovale and patent ductus arteriosus *B*, with atrial septal defect. *C*, with ventricular septal defect

28 Transposition of the Great Vessels

A. COMPLETE TRANSPOSITION

IF THE aorta arises from the right ventricle and the pulmonary artery from the left ventricle, *two separate circulatory systems* coexist. In one of them, which comprises the left side of the heart and the pulmonary circuit, oxygenated blood circulates. In the other, consisting of the right side of the heart and the systemic circuit, the circulating blood is venous. During fetal life, this malformation does not imply any markedly pathologic circulation, because the blood in the circulatory systems is *effectively mixed* through the foramen ovale and the ductus arteriosus. The gas exchange takes place in the placenta, which belongs to the systemic circulation. The only deviation from the normal course is that the more oxygenated blood from the inferior vena cava—of which a large proportion normally passes, via the foramen ovale and the left side of the heart, to the upper part of the body—returns instead to the lower part of the body through the left side of the heart, the pulmonary artery and the ductus arteriosus.

Immediately after birth, once the placental *circulation has ceased and the* pulmonary circulation is established, a pathologic circulation arises If there is no communication between the two systems, death ensues. The larger the communication, the greater chance the patient has of surviving. For the blood is well oxygenated in the lungs, but the oxygenated blood has difficulty in reaching the systemic circuit

In the presence of an extremely large ventricular septal defect, when the septum *is entirely lacking or is so poorly developed* that the blood from the ventricles is completely mixed, the great vessels are frequently transposed This often applies in tricuspid or mitral atresia. In these cases with a single functioning ventricle, the relative position of the great vessels is of no importance whatever for the hemodynamics, consequently, they should not be classified as cases of transposition

The communication between the two circulatory systems usually consists of a patent foramen ovale or atrial septal defect, patent ductus arteriosus, ventricular septal defect, or a combination of these anomalies Some examples of the different types of circulation are shown in Figure 680. A less common communication is by means of an abnormal venous return, the opening either of a systemic vein into the left atrium or of a pulmonary vein into the right atrium (as a rule, via a systemic vein). This is hard to establish clinically and is easily *overlooked at autopsy.* It has also been maintained that bronchial arteries and veins are able to convey a large proportion of the flow between the two systems (27, 216).

The importance of these associated malformations is evident from the survey of

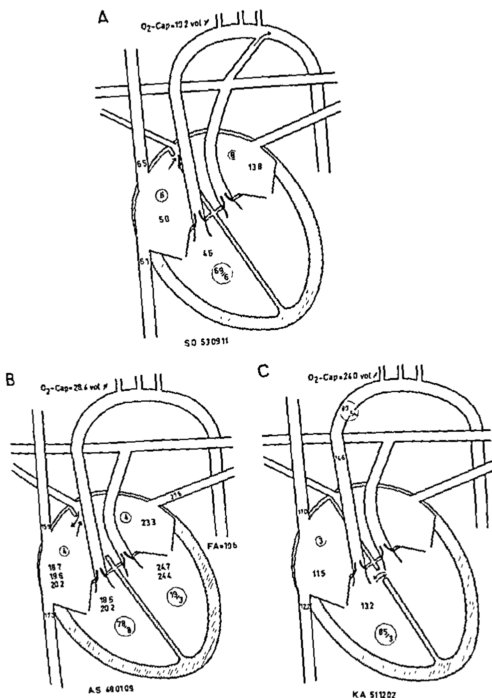


Fig. 680.—Examples of the hemodynamics in complete transposition of the great vessels *A*, with patent foramen ovale and patent ductus arteriosus. *B*, with atrial septal defect. C, with ventricular septal defect

123 cases reported by Hanlon and Blalock (315). The patients with a ventricular septal defect lived the longest, the mean figure being 4 years and 1 month, whereas the mean survival time for the whole series was 19 months. Complicating pulmonary stenosis is often present and alters the clinical syndrome appreciably (47).

Transposition of the great vessels is a relatively common malformation. Keith *et al.* (381) assembled 62 of their own cases during a 10 year period, but only 44 were typical, uncomplicated cases. Astley and Parsons (27) have given an account of 16 cases. Nadas (502) found 36 cases in a series of 577 patients with congenital heart disease.

Two autopsy cases are demonstrated in Figures 681 and 682.

Our series, consisting of 15 cases (10 boys and five girls), has been divided into the following three groups:

1. With a patent foramen ovale and patent ductus arteriosus (three cases).
2. With an atrial septal defect (three cases).
3. With a ventricular septal defect (nine cases)

In six of the nine cases in group 3 there were additional malformations i.e., atrial septal defect (one case), patent ductus arteriosus, atrial septal defect, and atresia of the left main branch of the pulmonary artery (one case), patent foramen ovale and valvular pulmonary stenosis (one case), and pulmonary stenosis (three cases). The age ranged from 2 weeks to 14 years.

CLINICAL FEATURES

Since in this malformation the circulation is practically normal during fetal life but exceedingly deficient postnatally, marked signs and symptoms are evident from birth onward The decreased ability of the oxygenated blood to pass into the systemic circulation results in severe cyanosis In order to maintain the oxygen transport, the cardiac output must be large. The heart volume increases rapidly from birth (see Fig. 692a).

Cyanosis is the most prominent symptom. It was present from birth in all of our cases except two, in which it was first observed at 1 month of age but subsequently increased rapidly in intensity. It has been stated that in cases with a patent foramen ovale and a patent ductus, cyanosis may be less severe in the lower half of the body thus indicating that the blood is shunted from the pulmonary to the systemic circuit through the patent ductus (27, 650). We did not find this to apply in our cases. Clubbing of the fingers had not developed in the children less than 4 months old. None of the three children who had started to walk was a squatter.

The physical development was retarded, and the older children were severely disabled. Three children had spells of unconsciousness with convulsions. Dyspnea during exertion or even at rest was often seen

The prognosis was poorest in the patients with only a patent foramen ovale and a patent ductus All of them died during the first year of life. Cases have, however, been known to survive until adult age (544). In our experience, the presence of a large atrial septal defect seems to be as favorable as a ventricular septal defect. Long survival in cases with only an associated atrial septal defect has, in fact, been reported by Rossi (569)

PHYSICAL SIGNS

The most characteristic cardiac finding is a greatly accentuated second sound over the pulmonary area. It is strikingly pure and presumably is to be ascribed to the closure of the aortic cusps alone The pulmonary orifice lies too far dorsally for the closure of its cusps to be heard or recorded

The murmurs may vary in nature, depending to some extent on the type of associated malformation A loud pansystolic murmur was present in seven of the nine patients with ventricular septal defect, but four of them also had pulmonary stenosis. A loud systolic murmur was also found in one patient with a patent foramen ovale and patent ductus, but the other patients

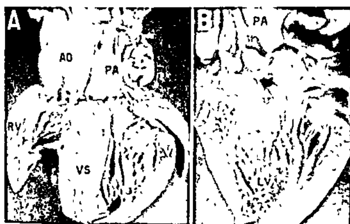


Fig 681 — ... the sam... membra... pulmon... artery), VS, ventricular septum ... entricles are of ... ow in B) in the ... ventricles, PA,

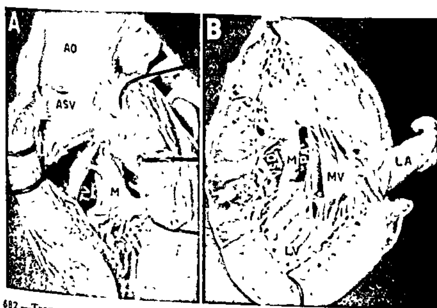


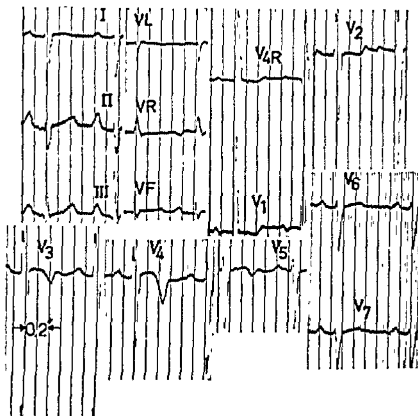
Fig 682 —Transposition ... ventricle and large left vent... illary muscles, ... part of the vent... cle and the pu... atrium, LV and... nary artery, TV ... ular septal defect

presented only a faint murmur or none at all. Lichtenstein and Mannheimer (435), Keith *et al.* (382), and Anderson and Adams (13) found that a systolic murmur was particularly characteristic of cases with an associated ventricular septal defect.

Electrocardiography

The right ventricle must work against the systemic pressure and with an increased blood flow Consequently, pronounced right ventricular hypertrophy is a constant finding in transposition of the great vessels. It was found in our 13 cases in which chest leads were used.

Since the flow through the left ventricle is also increased, signs of left ventricular hypertrophy are sometimes present as well This applied in one case (K.Ö. 521130), in which the communication between the circulatory systems consisted of a patent foramen ovale and patent ductus. The pressure must then be higher in the left ventricle than in the right. If there is a large atrial septal defect with mixing of the blood in both directions, the pressure in the left ventricle may fall to the level of the normal pulmonary artery pressure. This could be established in case A S. 480109, and the ECG therefore showed enormous dominance of the right ventricle (Fig. 683). Pathologic P waves (broad, notched, or peaked) were present in eight cases.



septal
/3.

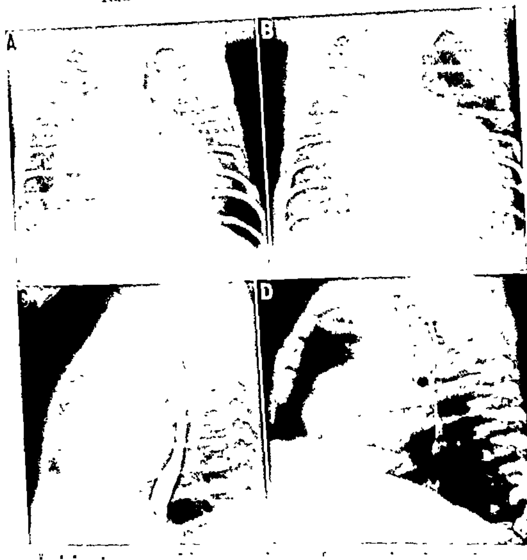
Roentgenologic Examination

The roentgenologic findings in transposition of the great vessels are usually characteristic. Taussig (649) was the first to describe them in detail. Further variants have since been reported (5, 27, 150).

The characteristic features are:

A change in the relative position of the aorta and pulmonary artery, whereby the latter is visualized very incompletely or not at all

Dilatation of the vessel branches centrally



and peripherally in the lungs, as an expression of an increased pulmonary circulation

Enlargement of the ventricles and frequently of both atria

All of our 15 patients underwent roentgenologic examination and 11 of them angiocardiographic examination as well.

In transposition of the great vessels, the aorta lies anteriorly and the pulmonary artery runs behind it, therefore, in the frontal view, these vessels are superimposed. As a rule, the aortic arch can[...] be identified In most cases, this is due [...] the fact that the aorta runs in a wide cu[...] close to the midline, more or less sagitta[...] In other cases, the cause is a markedly h[...] zontal course, so that the aortic root [...] extremely far to the left and the aortic a[...] medially (27) The mediastinum is th[...] fore seen to be narrow. We observed [...] most distinctly in infants (Figs. 684–6[...] but it was also fairly characteri[...]

presented only a faint murmur or none at all. Lichtenstein and Mannheimer (435), Keith *et al.* (382), and Anderson and Adams (13) found that a systolic murmur was particularly characteristic of cases with an associated ventricular septal defect.

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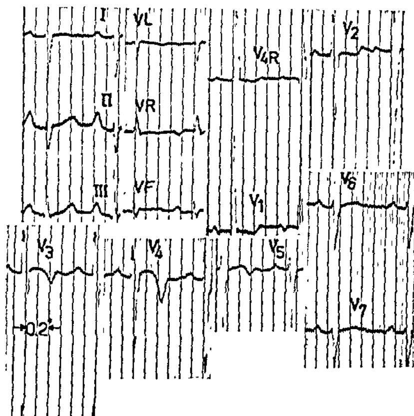


Fig. 683.—Electrocardiograms in complete transposition of the great vessels and atrial septal defect. *Boy, aged 6 (A S. 480109) Pressure in right ventricle 78/8 mm Hg, in left ventricle 19/3*

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The roentgenologic findings in transposition of the great vessels are usually characteristic Taussig (649) was the first to describe them in detail. Further variants have since been reported (5, 27, 150)

The characteristic features are

A change in the relative position of the aorta and pulmonary artery, whereby the latter is visualized very incompletely or not at all

older children (Figs 687 and 688). Keith *et al* (381) have pointed out that the narrowness of the mediastinum is most striking at birth, before the heart occupies a larger part of the space in it, owing to its increased volume In our cases, the dilated pulmonary artery was only exceptionally depicted in the frontal view (Fig. 689).

The thymus may occasionally cause difficulties in the evaluation, but it was not cending aorta can seldom be observed in the lateral view.

The essential cause of the dilatation of the central and peripheral pulmonary vessels is the increased flow, but the raised pressure in the pulmonary circulation must also be a contributory factor. The central vessels of the lungs often exhibit distinct pulsations. Reduced vascularity of the lungs was present in three cases in which there

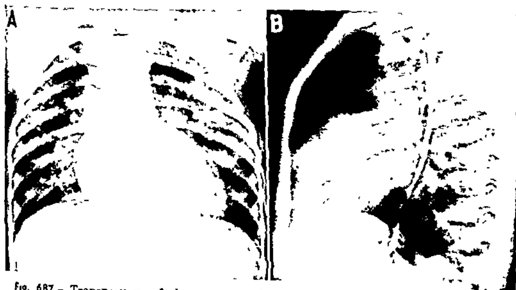


Fig. 687.—*Transposition of the great vessels and atrial septal defect. Boy, aged 6 (A S 30109), see Figure 695 Considerable hypertrophy ... dicle is not distinctly narrower than normally, but ... visualized Greatly increased vascularity of lungs*

sible in our cases As a rule, the variations the course of the aorta are greater than that of the pulmonary artery, and in a ss common variant the aorta may ascend a wide curve to the left in the mediastinum (27, 150). In many such cases, corected transposition of the great vessels resumably is present

In oblique projections, the width of the mediastinum is usually increased This applied in our cases, but it was somewhat difficult to estimate The width should be maximal in the lateral projection, but it is hard to judge owing to the fact that the great vessels are poorly visualized in this projection The ventral course of the

was associated pulmonary stenosis or atresia (Figs 688 and 690).

Enlargement and hypertrophy of the right ventricle are typical features and can almost invariably be demonstrated roentgenologically (Figs 685–687). The left ventricle, on the contrary, is difficult to evaluate because of the coincident right ventricular enlargement. The shape of the ventricular region is generally characteristic, it has a long, laterally bulging segment and a distinctly angulated apical portion, which is often separate from the diaphragmatic dome and is directed downward and outward (Figs. 684–687). No marked concavity at the

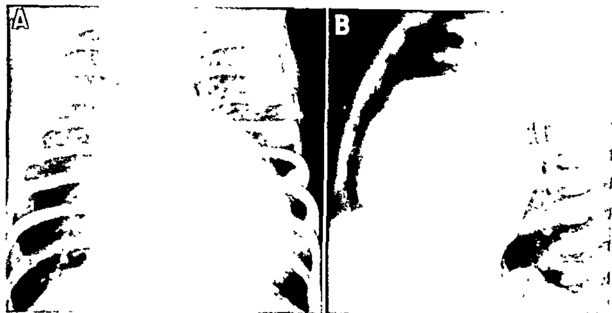


Fig. 685.—Transposition of the great vessels and small atrial septal defect. Boy, aged 3 months (B.E 520821). Considerable increase in heart volume, with enlargement of all chambers, particularly the right atrium and ventricle. Narrow vascular pedicle. Neither the aorta nor the main trunk of the pulmonary artery can be visualized. Greatly increased vascularity of lungs.

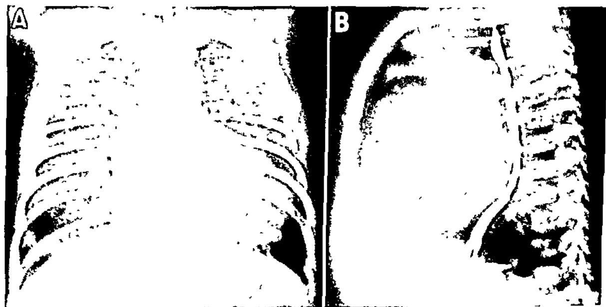


Fig. 686.—Transposition of the great vessels, patent foramen ovale, and patent ductus arteriosus Girl, aged 4 months (K Ö 521130) Considerable enlargement of heart, especially the right ventricle and left atrium Vascular pedicle is narrow, but somewhat broader than in the preceding cases, as in those cases, no aortic arch segment is visible Main trunk of the pulmonary artery cannot be visualized Greatly increased vascularity of lungs



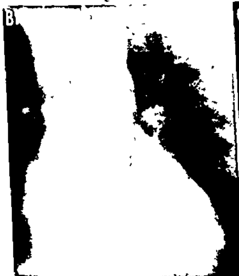




Fig 690.—Transposition of the great vessels Girl, aged 14 (M S 410726) Reduced blood volume in pulmonary circulation, with irregular, reticular vascular markings in both lungs, as in collateral circuit Considerable dilatation of aorta Thus it is seen distinctly in *B* that the left outline both of the aortic arch and that of descending aorta lie fairly far to the left. The main trunk and main branches of the pulmonary artery are indistinct, and the impression in the esophagus is lengthy

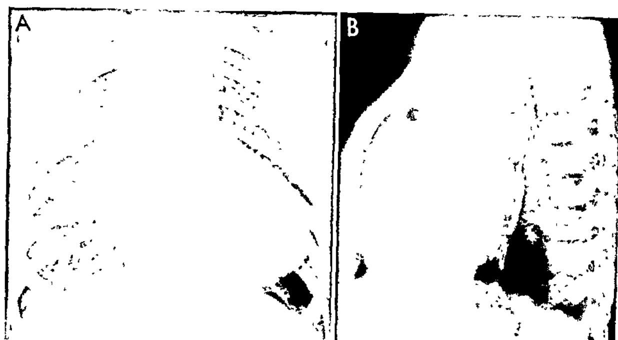


Fig. 688.—Transposition of the great vessels. Girl, aged 3 (E P 520605) Great hypertrophy of right ventricle, with upturning of apex and bulging of anterior surface The main trunk of the pulmonary artery cannot be delimited. Upper mediastinum narrow. Decreased blood volume in pulmonary circulation

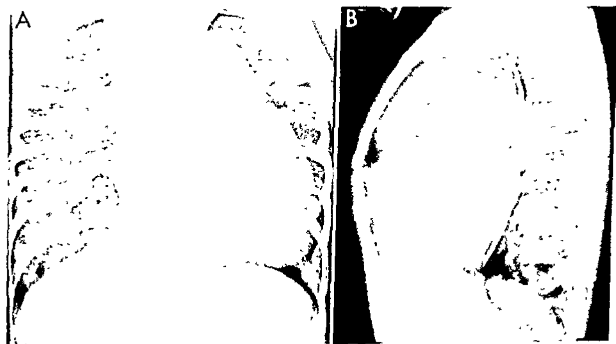


Fig. 689 —Transposition of the great vessels Boy, aged 3 (H E 510828) Pulmonary artery wide and runs in abnormal direction far to left Hypertrophy of right ventricle, with forward bulging of anterior thoracic wall Increased blood volume in pulmonary circulation and enlargement of left atrium

An enlarged right atrium was observed in more than half the cases. In several the enlargement was extremely marked (Figs. 684, 685, and 691). We were not able to establish definitely a feature demonstrated by Taussig (649), namely, rhythmic variations in the volume of the right atrium when the communication between the two circuits consists of a patent foramen ovale and patent ductus.

Enlargement of the left atrium was present in nine cases; in seven of them the dilatation was fairly great or considerable (Figs. 686 and 691) This was confirmed in the three autopsy cases The dilatation is a manifestation of the increased blood volume of the lungs and appears to be a roentgenologic feature as typical as the enlargement of the right atrium.

The heart is almost invariably enlarged The heart volume was increased in all of our cases, in seven of them to twice the normal volume or more Taussig (649) has shown that the heart rapidly becomes enlarged postnatally This is illustrated by two of our cases (Figs 684 and 692)

The variations in the roentgenologic appearance depend chiefly on the volume of the shunts and, to a lesser degree, on their localization. Thus, the appearance was the same in the cases with associated patent foramen ovale and patent ductus (Figs. 684 and 686) and in one case with a small atrial septal defect only (Fig. 685). The increased blood volume of the lungs, which is essentially compensatory, is an expression of the inadequate communication between the right and left side of the heart. All the chambers of the heart were enlarged in these cases. In another case with an atrial septal defect, in which the shunt passed through a probably wide communication, the appearance differed from the foregoing. Thus, the vascularity of the lungs was only slightly increased, the left atrium was not dilated, and the heart was far less enlarged.

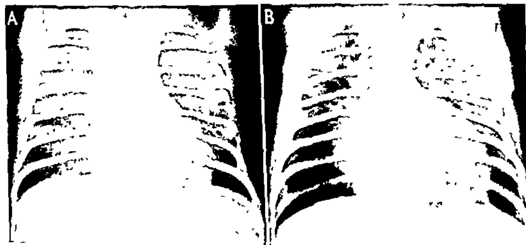


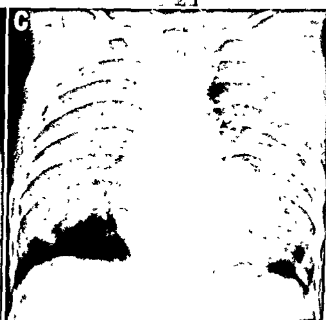
Fig 692. ... birth ... ance ... hear ... of c... transposition ... already has the appearance typical

Four cases with an associated ventricular septal defect also presented, in all essentials, the same typical appearance as that first described In one of them (Fig. 693), not investigated by angiocardiography, autopsy revealed in addition mild valvular pulmonary stenosis.

From a differential diagnostic viewpoint it must be ...







F[illegible] [illegible]71 [illegible]
5112 [illegible]
only [illegible]
vascularity of lungs.

appearance in transposition of the great vessels shows many similarities to that in pulmonary atresia with a wide patent ductus. In both conditions increased vascularity of the lungs is typical, despite the fact that the main trunk of the pulmonary artery cannot be identified Moreover, the ventricles are enlarged, and the left atrium may be dilated and the heart volume increased In pulmonary atresia, however, the aorta is usually greatly dilated and the aortic arch is prominent (cf Fig 249, p.

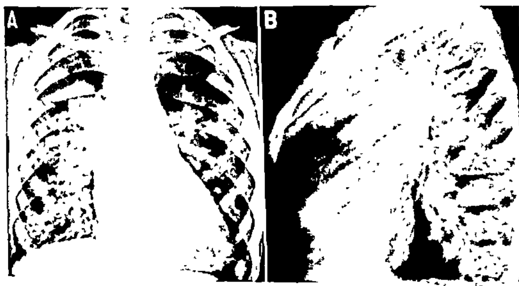


Fig. 693.—Transpositior . . septal defect Boy, aged 12 right atrium, pulmonary ar monary vessels, greatly incr ... ...atation of left atrium

269) There may be identical findings in true persistent truncus arteriosus (cf. Fig. 306, p 332)

Cardiac Catheterization

The clinical features, the ECG and the roentgenologic appearance are usually so characteristic as to establish the diagnosis The typical features are severe cyanosis with an early onset, considerable cardiac enlargement and an increased pulmonary flow The nature of the ... mations is ... lish withc

cardiac catheterization and angiocardiography are required, sometimes on several occasions, with injection into both circulatory systems.

The anterior position of the aorta can be established by fluoroscopy during catheterization. It is seldom possible to demonstrate transposition of the pulmonary artery in this way.

Cardiac catheterization is of diagnostic value only if pressure recordings and blood samples can be obtained from all four chambers of the heart and from the great vessels. This was, however, possible in only two of our 11 cases in which catheterization was performed. The blood in the aorta has a lower oxygen content than that in the pulmonary artery. Evaluation of the volume of the shunts is uncertain, since they vary greatly from moment to moment (130).

Figure 680 shows examples of the findings on cardiac catheterization in transposition of the great vessels combined ...

A

B

AO

PA

LV

C

D

RV

LV



Fig. 692b.—Same case as in Figure 692a Pulmonary artery (*PA*) somewhat wider than aorta (*AO*) Small atrial septal defect (left arrow in *E*) at site of foramen ovale Right arrow in *E* points to tricuspid orifice. Right ventricle (*RV*) considerably more thick walled than left (*LV*). cf. *RV* and *LV* (left ventricle) in *C* *RA*, right atrium.

resistance in the two vascular systems (see p 339) The relation between the resistance of the circulations probably varies considerably with the respiration In an interatrial shunt as well, the respiration is an important factor. As was pointed out in the discussion of the hemodynamics in atrial septal defect, respiration causes greater variations in the pressure in the left atrium than in that in the right (p. 439).

The variations in the shunt from one moment to the next could be demonstrated in case A S 480109, since many samples were taken from the different chambers of the heart The oxygen content of samples from the right atrium ranged from 18 5 to 20 2 volumes per cent, and equally large differences were found in samples from the right ventricle. An analysis of the pressure curves from both atria provided an explanation of this variation (Fig 694). Unfortunately, we were unable to take simultaneous pressure tracings from both atria, but the recordings were made under similar conditions The most outstanding difference was that respiration was associated with considerably greater variations in the left atrium than in the right Consequently, the pressure was lower in the left atrium than in the right during inspiration and higher during expiration

There is also a possibility of a change in the direction of the interatrial shunt during each cardiac cycle In the case in question, the right ventricle was a high-pressure chamber, whereas the left ventricle was a low-pressure chamber In direct contrast to the normal conditions, the right ventricle offered greater resistance to diastolic filling than did the left ventricle During ventricular diastole, the blood may be shunted from the right atrium to the left atrium. A rapid fall in pressure on opening of the mitral valve is also seen on the pressure curve from the left atrium, it is considerably more pronounced than in cases with normal pressure in the left ventricle On the other hand, the v wave is, as normally, higher in the left atrium This is due to the fact that the left atrium, because of its structure, is less distensible than the right It should therefore be possible for a left to right shunt to take place during this phase (cf. p. 440).

ANGIOCARDIOGRAPHY

In our series, 11 of the patients underwent an angiocardiographic examination. In seven cases the contrast medium was injected into the right ventricle, in three into the left ventricle, and in one by the intravenous route. A complementary examination with injection into the left ventricle was made on a later occasion in one of the cases investigated by right ventricular injection.

As far as the choice of projection is concerned, many authors recommend the left anterior oblique view (270). However, for the same reasons as in other forms of transposition of the aorta (p. 820), the relation of this vessel to the right ventricle is visualized best in the true lateral view. In our investigations, the films were exposed synchronously in the frontal and lateral projections in every case.

The communication between the right and the left side of the heart is usually small. Consequently, it is seldom possible to make a complete analysis of the anatomy of both circulatory systems on a single examination. This applies irrespective of whether intravenous injection is used or a selective procedure with injection of the contrast medium into one of the ventricles or atria Each system must be visualized separately.

Transposition of the aorta is best established selectively, by injection of the contrast medium into the right ventricle. The aorta arises entirely from the infundibulum The site of the aortic orifice does not, however, correspond exactly to the normal position of the pulmonary orifice, since the infundibulum is nearly always directed obliquely upward to the right (Figs 695 and 699). The aortic orifice may occasionally lie far to the left. A feature common to all the cases is that the ascending aorta lies ventrally and joins the descending aorta in a wide curve The aorta ...

to right intracardiac shunt. The catheter *did not* pass into the aorta and arterial *puncture* was unsuccessful. It should be *possible* to demonstrate the reversed flow through the patent ductus by simultaneous puncture of the brachial and the femoral artery, since the oxygen saturation should be higher in the latter. In one case the right to left interatrial shunt could be established.

In the presence of an atrial septal defect (Fig. 680, *B*), an interatrial shunt in both *directions* was demonstrated. No other shunts were present. The pressure in the left ventricle corresponded to the normal pulmonary artery pressure, whereas that in the right ventricle was systemic. These *findings are decisive for the diagnosis.*

If the atrial septum is intact and the ventricular septal defect is small, it usually is not possible to advance the catheter into the left side of the heart. Therefore, in a case of this kind, only a left *to* right interventricular shunt could be demonstrated (Fig. 680, C). The abnormal site of the aorta was demonstrated on fluoroscopy, by means of passage of the catheter from the right ventricular into the aorta, which was seen to lie far ventrally. The fact that the oxygen content of the aorta is higher than *that of the right ventricle is not proof of* patency of the ductus, since this often applies in uncomplicated ventricular septal defect.

In a case with a large ventricular septal defect, the catheter entered the left ventricle and the pulmonary artery. In another case with both a ventricular and an atrial septal defect, the left side was entered through the interatrial communication. A shunt was present at both the atrial and the ventricular level, but the oxygen content of the pulmonary artery was much higher *than that of the aorta.*

The way in which the blood is shunted between the circulatory systems has been the subject of much speculation The direction of the circuit has been discussed by Becker and Brill (47) on the basis of the autopsy findings in three cases. In their opinion, when both the foramen ovale and the ductus arteriosus are patent, the blood flows from the right atrium to the left and from the pulmonary artery to the aorta. They also state that, with an associated atrial septal defect, *the interatrial shunt is* mixed, and with a ventricular septal defect the direction is from left to right during systole, and vice versa during diastole. Campbell *et al.* (130) have expressed the view that, even if the shunt must be equally large in both directions, it can—during various short periods—flow mainly in one or the other direction. Taussig (650) stresses that, with a patent foramen ovale combined with a patent ductus, the interatrial shunt can be directed only from right to left, provided the valve covers the opening *and impedes the flow in the opposite direc*tion. The shunt through the ductus would pass from the pulmonary artery to the aorta. The pressure then decreases in the left atrium and increases in the right, until the foramen ovale opens and the blood is shunted into the left atrium. She was able to observe at fluoroscopy how the right atrium increased successively in size and then collapsed suddenly. This rhythmic variation in volume was independent of the cardiac rhythm. In a similar way, the shunt through a ventricular septal defect would *change direction periodically*, by means of a shunt in one direction causing a rise in pressure in one of the systems, after which the shunt would be reversed. Campbell *et al.* (130) were, however, unable to observe any variations in pressure either in the ventricles or in the atria

Astley and Parsons (27) have stated that *the shunt through the ventricular septal* defect goes from right to left and the reflow through the bronchial veins. With an atrial septal defect, they consider that the direction is from left to right, the reflow taking place through the bronchial arteries

As is always the case with a large ventricular septal defect, the pressure in the *two ventricles is equilibrated*, as is the systolic pressure in the two arterial systems, provided neither pulmonary nor aortic stenosis is present The volume and direction of the shunt are determined by the total

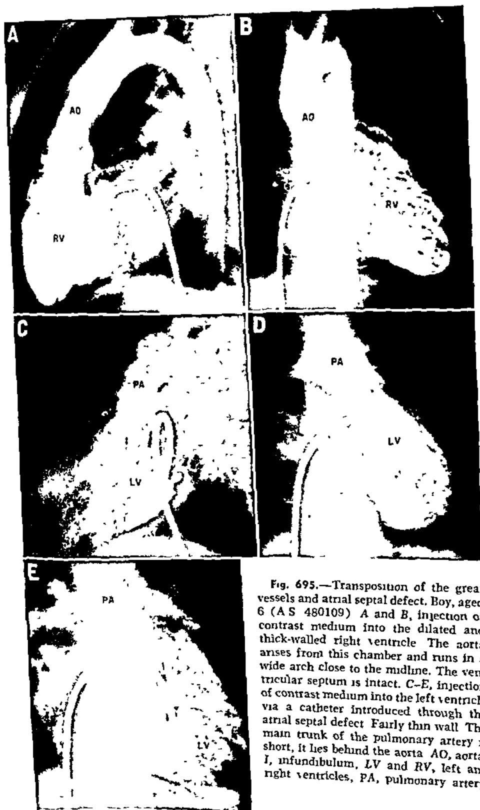


Fig. 695.—Transposition of the great vessels and atrial septal defect. Boy, aged 6 (A S 480109) *A* and *B*, injection of contrast medium into the dilated and thick-walled right ventricle The aorta arises from this chamber and runs in a wide arch close to the midline. The ventricular septum is intact. *C–E*, injection of contrast medium into the left ventricle via a catheter introduced through the atrial septal defect Fairly thin wall The main trunk of the pulmonary artery is short, it lies behind the aorta *AO*, aorta, *I*, infundibulum, *LV* and *RV*, left and right ventricles, *PA*, pulmonary artery

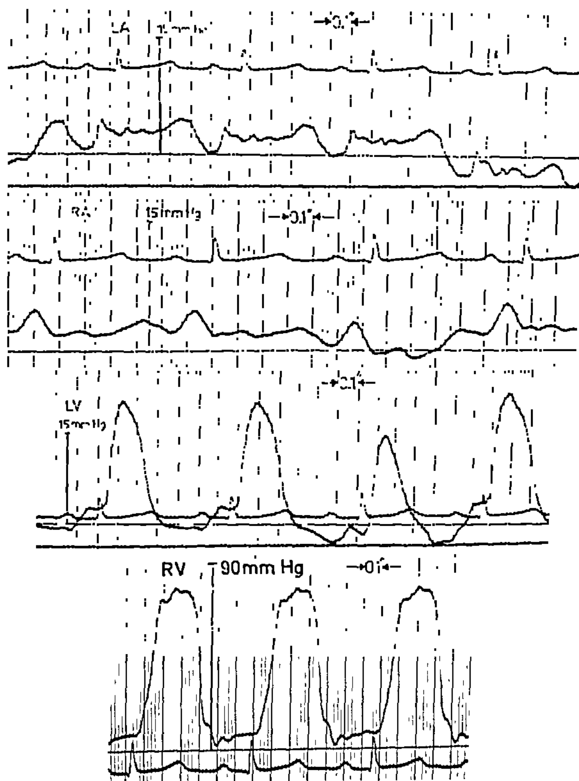


Fig. 694.—Complete transposition of the great vessels and atrial septal defect. Boy, aged 6 … ventricles show that the right ven- … orresponding to normal pulmonary … cle results in unusually rapid *fall in* pressure in the left atrium at the opening of the mitral valve, this permits a right to left interatrial shunt. Since the left atrium is less distensible than the right, pressure is higher in the former during ventricular systole, and a left to right interatrial shunt occurs The greater respiratory variations in pressure in the left atrium also permit a periodic change in direction of the shunt.

cardiographic examination. A patent ductus was distinctly depicted in one case (Fig. 697) A surgical anastomosis between the right subclavian artery and the pulmonary artery is shown in Fig 698

The left side of the heart can be visualized selectively if the catheter can be passed through a patent foramen ovale or an atrial or a ventricular septal defect The contrast medium may be injected into the atrium or the ventricle This procedure was used as a complementary examination in one of our cases in order to obtain a more complete picture of the anatomy (Fig 695) The pulmonary artery, which was short, ran backward to the right The ventricular septum was found to be intact, and the communication was in the form of an atrial septal defect (Fig. 680, *B*). The left ventricle was smaller than the right ventricle, the wall of which was hypertrophic, whereas that of the former was thin. These observations are thus in good agreement with the pressure conditions in recordings from the ventricles (p. 788).

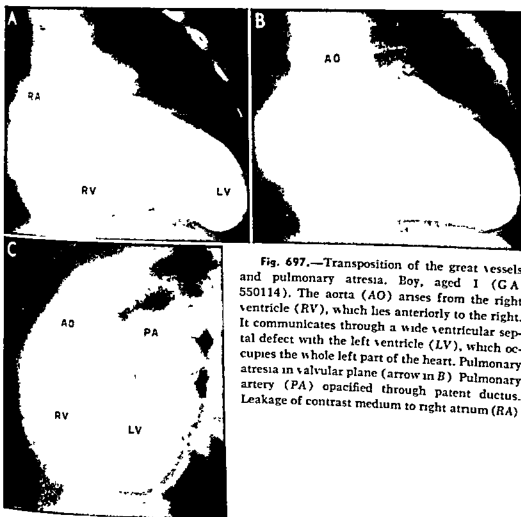


Fig. 697.—Transposition of the great vessels and pulmonary atresia. Boy, aged 1 (G A 550114). The aorta (*AO*) arises from the right ventricle (*RV*), which lies anteriorly to the right. It communicates through a wide ventricular septal defect with the left ventricle (*LV*), which occupies the whole left part of the heart. Pulmonary atresia in valvular plane (arrow in *B*) Pulmonary artery (*PA*) opacified through patent ductus. Leakage of contrast medium to right atrium (*RA*)

B. INCOMPLETE TRANSPOSITION (TAUSSIG-BING COMPLEX)

Just as the position of the aortic root may vary from normal to complete transposition, the pulmonary artery root may be

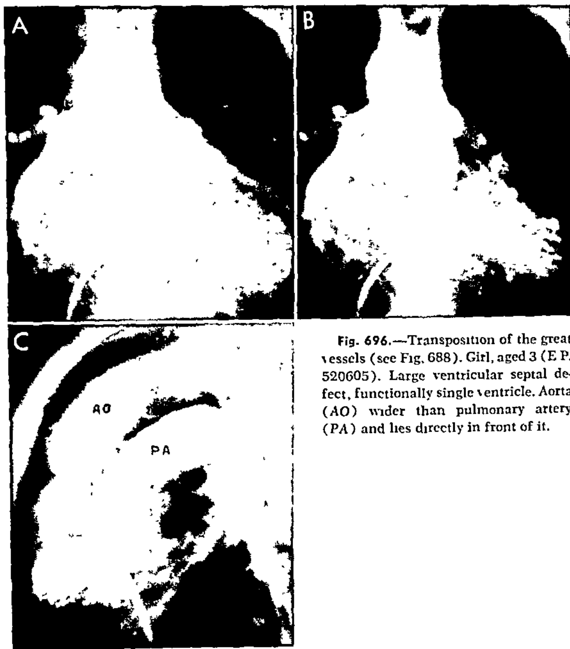


Fig. 696.—Transposition of the great vessels (see Fig. 688). Girl, aged 3 (E P. 520605). Large ventricular septal defect, functionally single ventricle. Aorta (AO) wider than pulmonary artery (PA) and lies directly in front of it.

width was seen with associated pulmonary stenosis or atresia (Figs 696-698).

If the ventricles communicate through a sufficiently wide defect, this can be depicted by injection of the contrast medium into the right ventricle. Both this ventricle and the aorta, as well as the left ventricle and pulmonary artery, are then visualized simultaneously (Figs. 696, 699, and 700). The right ventricle and aorta can be demonstrated in a corresponding way after injection of contrast medium into the left ventricle (Figs 697, 698, and 701) Stenosis or atresia of the pulmonary orifice was disclosed by these means in three cases. Stenosis and atresia are illustrated in Figures 697 and 698

An interatrial communication can be demonstrated by intravenous injection of contrast medium (158) or by selective injection into the right atrium

In two cases in which a patent ductus had been diagnosed clinically, it could not be demonstrated with certainty on angio-

cardiographic examination. A patent ductus was distinctly depicted in one case (Fig. 697) A surgical anastomosis between the right subclavian artery and the pulmonary artery is shown in Fig 698.

The left side of the heart can be visualized selectively if the catheter can be passed through a patent foramen ovale or an atrial or a ventricular septal defect The contrast medium may be injected into the atrium or the ventricle This procedure was used as a complementary examination in one of our cases in order to obtain a more complete picture of the anatomy (Fig 695) The pulmonary artery which was short, ran backward to the right The ventricular septum was found to be intact, and the communication was in the form of an atrial septal defect (Fig. 680, B). The left ventricle was smaller than the right ventricle, the wall of which was hypertrophic, whereas that of the former was thin These observations are thus in good agreement with the pressure conditions in recordings from the ventricles (p. 788).

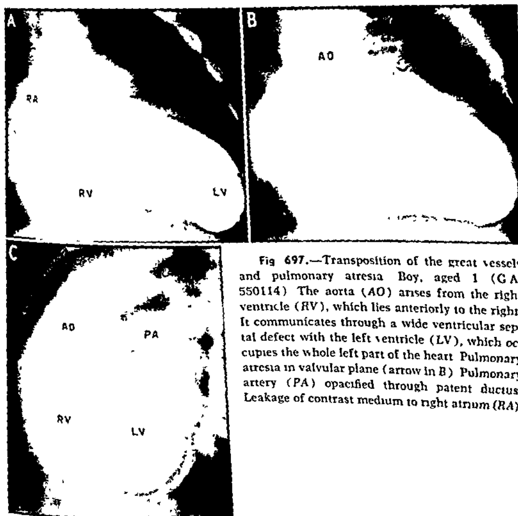


Fig 697.—Transposition of the great vessels and pulmonary atresia Boy, aged 1 (G A 550114) The aorta (*AO*) arises from the right ventricle (*RV*), which lies anteriorly to the right. It communicates through a wide ventricular septal defect with the left ventricle (*LV*), which occupies the whole left part of the heart Pulmonary atresia in valvular plane (arrow in *B*) Pulmonary artery (*PA*) opacified through patent ductus Leakage of contrast medium to right atrium (*RA*)

B INCOMPLETE TRANSPOSITION (TAUSSIG-BING COMPLEX)

Just as the position of the aortic root may vary from normal to complete transposition, the pulmonary artery root may be

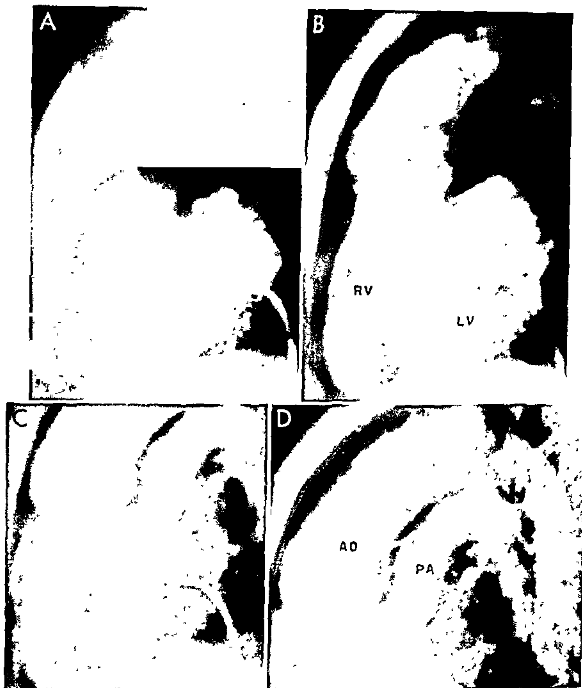


Fig. 698.—Transposition of the great vessels (see Fig 690). Girl, aged 14 (M.S 410726) The wide aorta (*AO*) lies directly in front of the narrow pulmonary artery (*PA*) Both ventricles are well developed, the aorta takes its origin from the right (*RV*) and the pulmonary artery from the left (*LV*) Stenosis at origin of pulmonary artery Large ventricular septal defect. Blalock-Taussig operation performed earlier, with anastomosis between right subclavian artery (*RSA*) and right main branch of the pulmonary artery (*continued*)



Fig. 698 (*cont.*)



Fig. 698.—Transposition of the great vessels (see Fig 690) Girl, aged 14 (M S 410726). The wide aorta (*AO*) lies directly in front of the narrow pulmonary artery (*PA*) Both ventricles are well developed, the aorta takes its origin from the right (*RV*) and the pulmonary artery from the left (*LV*) Stenosis at origin of pulmonary artery Large ventricular septal defect Blalock-Taussig operation performed earlier, with anastomosis between right subclavian artery (*RSA*) and right main branch of the pulmonary artery (*continued*)



Fig 700 —Transposition of the great vessels Boy, aged 6 (K J 490702) The right atrium

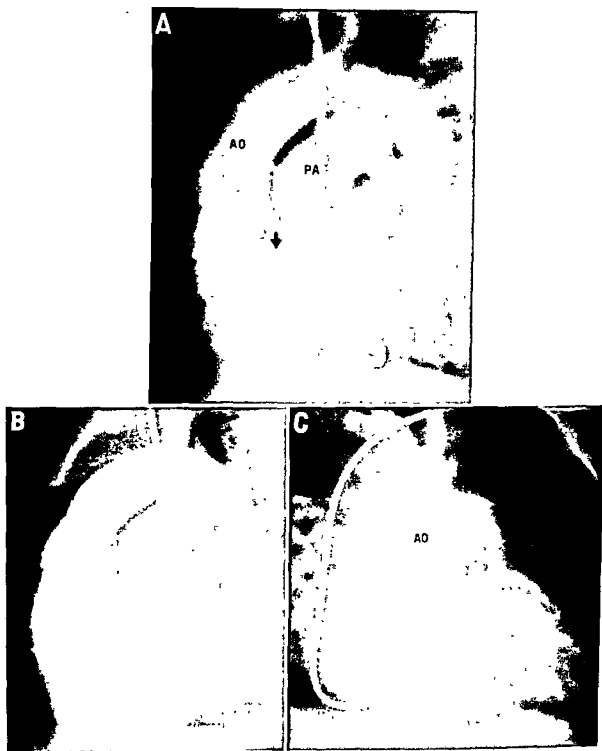


Fig. 699.—Transposition of the great vessels and ventricular septal defect Boy, aged 2 (K A 511202). Injection of contrast medium into the large right ventricle Wide septal defect (arrow

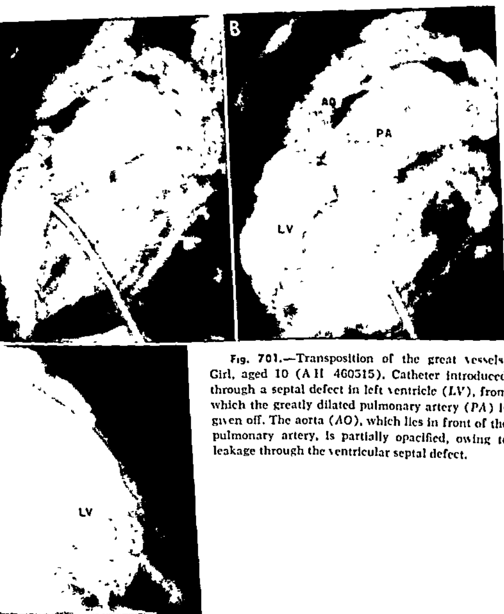


Fig. 701.—Transposition of the great vessels. Girl, aged 10 (A H 460515). Catheter introduced through a septal defect in left ventricle (*LV*), from which the greatly dilated pulmonary artery (*PA*) is given off. The aorta (*AO*), which lies in front of the pulmonary artery, is partially opacified, owing to leakage through the ventricular septal defect.

er-riding (107) or completely transposed. n incomplete transposition of the great essels, both are at an abnormal site, but ne of them is not completely transposed. n anatomic description of the malforma- on was given by Brown (107) as early as 939, whereas the characteristic complex vas first described in its entirety by Taussig nd Bing (652) and has been given their name It consists of "transposed aorta, a large pulmonary artery which arises primarily from the right ventricle and partially over-rides the ventricular septum, a high ventricular septal defect and a right ventricular hypertrophy." The aorta lies ventrally and to the right of the pulmonary artery. At autopsy, the latter appears on the exterior to be "approximately in its normal position," but is found on dissection to be over-riding At angiocardiography, we have

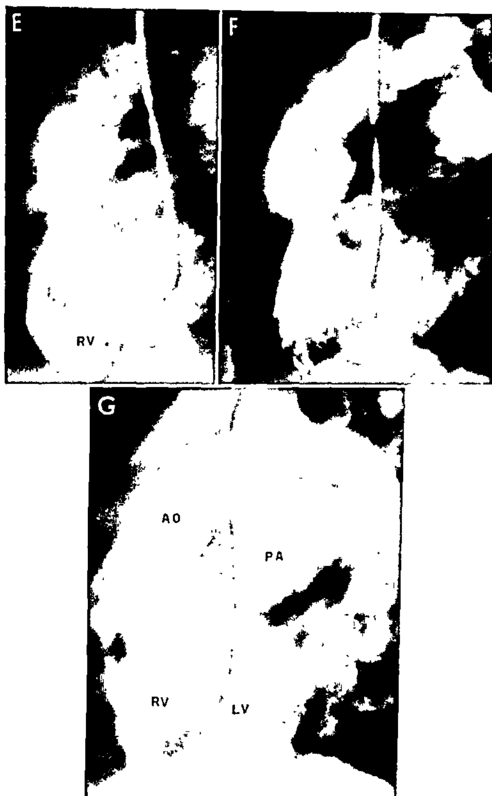


Fig. 700 (*cont.*)

performed, with end-to-end anastomosis between right subclavian artery and pulmonary branch to right upper lobe (*D*). Postoperatively, considerable dilatation of pulmonary branches (D_1) to upper lobe

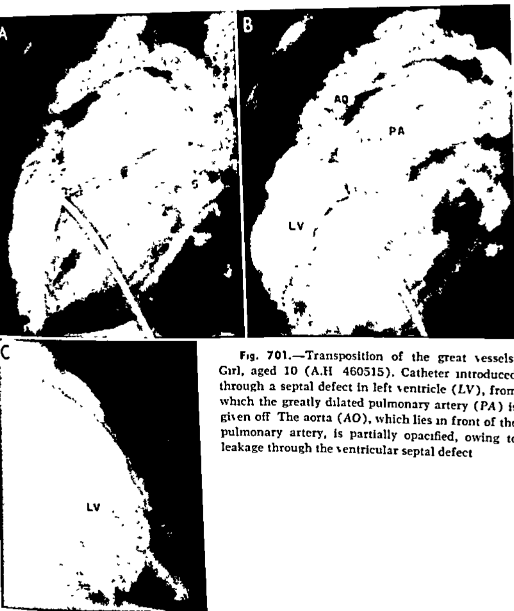


Fig. 701.—Transposition of the great vessels Girl, aged 10 (A.H 460515). Catheter introduced through a septal defect in left ventricle (*LV*), from which the greatly dilated pulmonary artery (*PA*) is given off The aorta (*AO*), which lies in front of the pulmonary artery, is partially opacified, owing to leakage through the ventricular septal defect

over-riding (107) or completely transposed In incomplete transposition of the great vessels, both are at an abnormal site, but one of them is not completely transposed An anatomic description of the malformation was given by Brown (107) as early as 1939, whereas the characteristic complex was first described in its entirety by Taussig and Bing (652) and has been given their name It consists of "transposed aorta, a large pulmonary artery which arises primarily from the right ventricle and partially over-rides the ventricular septum, a high ventricular septal defect and a right ventricular hypertrophy." The aorta lies ventrally and to the right of the pulmonary artery At autopsy, the latter appears on the exterior to be "approximately in its normal position," but is found on dissection to be over-riding. At angiocardiography, we have

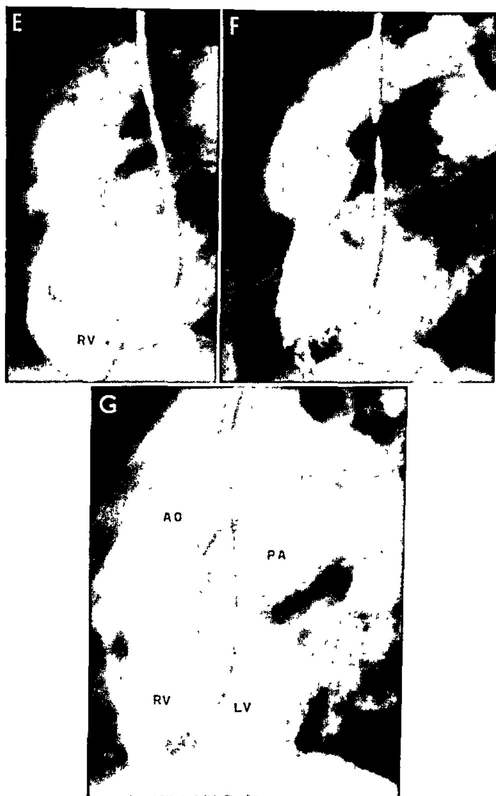


Fig. 700 (*cont*)

performed, with end-to-end anastomosis between right subclavian artery and pulmonary branch to right upper lobe (*D*). Postoperatively, considerable dilatation of pulmonary branches (D_1) to upper lobe

ROENTGENOLOGIC EXAMINATION

In three of the cases, the appearance was consistent with that described as characteristic of the Taussig-Bing complex (487, 652). There were considerable dilatation of the main trunk of the pulmonary artery and greatly dilated and pulsating central and perihilar vessels in the lungs, marked hypertrophy and enlargement of the right ventricle, and probable enlargement of the left ventricle as well. The right atrium was distinctly enlarged, and the left atrium slightly dilated. The peripheral vessels in the lungs were definitely dilated, although not in proportion to the central vessels In all essentials, the appearance was reminiscent of that in ventricular septal defect with pulmonary hypertension

Angiocardiography showed the main trunk of the pulmonary artery to take its origin more dorsally than normal. Consequently, on conventional roentgenologic examination in the right anterior oblique projection, the pulmonary artery should have been far less distinct than in the frontal view. This finding did, in fact, apply in all three cases However, this sign can scarcely be used with any great degree of certainty for the diagnosis, in view of the fact that similar variations in the [illegible] forms of heart disease with dilatation of the pulmonary artery

In the fourth case, the roentgenologic appearance was uncharacteristic, owing to the fact that the heart was rotated and displaced to the left, as an effect of widespread left-sided atelectasis of the lung.

Pulmonary stenosis, which has been observed by other authors as an associated malformation (663), was present in *none* of our cases

CARDIAC CATHETERIZATION

If the catheter passes into both the aorta and the pulmonary artery, the position of the great vessels can be determined on fluoroscopy, but their relation to the ven-

TABLE 30 —TAUSSIG-BING COMPLEX: RESULTS OF CARDIAC CATHETERIZATION IN 4 CASES*

Case	O_2 Content, vol %										O_2 Cap., vol %	Pressure, mm Hg									
	SVC	IVC	RA	RV	PV	LA	LV	PA	Aorta	Fem. A.		RA	RV		LA	LV		PA		Aorta	
												Mean	Syst	End-diast	Mean	Syst	End-diast.	Syst	Diast	Syst	Diast.
L K 530325	12.0	10.0	10.8	12.7	—	—	—	14.1	—	13.9	19.6	−3	95	7	—	—	—	97	49	—	—
U O 450908	14.1	14.9	14.9	16.3	—	—	21.5	—	17.1	—	27.6	−1	91	3	—	93	3	—	—	92	87
O E 520424	9.8	9.2	10.2	18.8 10.4	—	17.2	14.8	—	—	—	18.9	5	91	9	7	97	7	—	—	—	—
K L 520412	9.7	9.7	10.1	15.0	18.8	18.4	18.6	15.8	—	11.7	19.4	2	82	2	5	90	9	80	46	—	—

*For abbreviations see Table 1, p 119

found that although the pulmonary artery appears to be at the normal site in the *frontal projection, it lies dorsally in the* lateral projection.

Taussig and Bing have stressed that, clinically, this complex resembles Eisen*menger's complex. The most salient differ*ence is that cyanosis is present from birth, whereas in Eisenmenger's complex it usually becomes noticeable only at puberty.

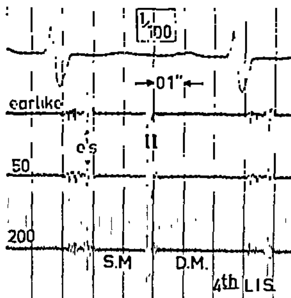


Fig 702.—Phonocardiogram in Taussig-Bing complex Boy, aged 9 (U O. 450908). Over the 4th left interspace *(4th L I S)*, an early systolic sound *(es)*, faint systolic murmur *(SM)*, accentuated second sound *(II)*, and faint protodiastolic murmur *(DM)* were recorded. Boxed figures denote degree of amplification, other figures denote standard frequencies of the filters

Our series contains five cases of this malformation. In one, the complex was combined with coarctation of the aorta with a fetal type of circulation through a patent ductus arteriosus. An account has been given of this case in connection with the former malformation (p 636). Several cases with associated coarctation of the aorta have been reported (6, 482, 487, 707).

The four other cases presented the following features

Boy, aged 11 months (L.K. 530325) —Congenital heart disease was diagnosed immediately after birth, on account of a murmur. He exhibited no other symptoms or signs until after 6 months of age. Cyanosis of the lips was then noticed when he cried, and later at rest as well He gained weight poorly and was unable to sit up at 11 months. He presented a slight precordial bulge, a systolic thrill over the apex, and pulsations over the precordium. An accentuated, split first sound (systolic click) and a short, loud systolic murmur were heard over the apex. A continuous murmur was audible over the first right interspace. The ECG showed combined left and right ventricular hypertrophy.

Boy, aged 9 years (U.O. 450908).—Cyanosis was first observed at 2 weeks of age. His physical development was retarded He was greatly disabled but was not a squatter. Examination disclosed marked, generalized cyanosis and clubbing of the fingers and toes The cardiac findings were characterized by a parasternal lift, an early, split first sound over the apex (systolic click), a greatly accentuated but pure second sound, much louder to the left of the sternum than to the right, an extremely short, faint systolic murmur over the second left interspace, and a long, faint diastolic murmur over the third interspace to the left of the sternum (Fig 702). The ECG showed pronounced right ventricular hypertrophy and tall P waves in the right precordial leads There was also transposition of the abdominal viscera

Boy, aged 4 years (O E 520424).—A systolic murmur was heard immediately after birth Cyanosis and dyspnea were noticed at 2 weeks of age He gained weight poorly. Both physical and mental development were markedly retarded At 4 years, he was unable to speak or walk and weighed only 11 4 kg. Examination showed severe cyanosis and marked clubbing of the fingers and toes The cardiac findings consisted of a precordial bulge, a parasternal lift, an accentuated but not split second sound, a faint early-systolic murmur over the pulmonary area and a mid-diastolic, short, low-frequency murmur over the apex The ECG disclosed marked right ventricular hypertrophy

Boy, aged 3 years (K L 520412) —*Congenital heart disease was diagnosed at* 2 months of age, because of a murmur Cyanosis was not observed before 6 months of age At 11 months he was able to walk, but was easily fatigued on exertion He presented severe cyanosis, clubbing of the fingers and toes, a parasternal lift, an accentuated, pure second sound, and a faint, early-systolic murmur over the pulmonary area The ECG showed marked right ventricular hypertrophy.

ROENTGENOLOGIC EXAMINATION

In three of the cases, the appearance was consistent with that described as characteristic of the Taussig-Bing complex (487, 652) There were considerable dilatation of the main trunk of the pulmonary artery and greatly dilated and pulsating central and perihilar vessels in the lungs, marked hypertrophy and enlargement of the right ventricle, and probable enlargement of the left ventricle as well. The right atrium was distinctly enlarged, and the left atrium slightly dilated The peripheral vessels in the lungs were definitely dilated, although not in proportion to the central vessels In all essentials, the appearance was reminiscent of that in ventricular septal defect with pulmonary hypertension

Angiocardiography showed the main trunk of the pulmonary artery to take its origin more dorsally than normal. Consequently, on conventional roentgenologic examination in the right anterior oblique projection, the pulmonary artery should have been far less distinct than in the frontal view This finding did, in fact, apply in all three cases However, this sign can scarcely be used with any great degree of certainty for the diagnosis, in view of the fact that similar variations in the prominence of the main trunk in different [illegible] obse[illegible] form[illegible] pulm[illegible] artery

In the fourth case, the roentgenologic appearance was uncharacteristic, owing to the fact that the heart was rotated and displaced to the left, as an effect of widespread left-sided atelectasis of the lung.

Pulmonary stenosis, which has been observed by other authors as an associated malformation (663), was present in none of our cases

CARDIAC CATHETERIZATION

If the catheter passes into both the aorta and the pulmonary artery, the position of the great vessels can be determined on fluoroscopy, but their relation to the ven-

TABLE 30 — TAUSSIG-BING COMPLEX. RESULTS OF CARDIAC CATHETERIZATION IN 4 CASES*

Case	O_2 Content vol %										O_2 Cap vol %	Pressure, mm Hg									
	SVC	IVC	RA	RV	PV	LA	LV	PA	Aorta	Fem A.		RA	RV		LA	LV		PA		Aorta	
												Mean	Syst	End-diast	Mean	Syst	End-diast.	Syst	Diast.	Syst	Diast.
I K 530325	12.0	10.0	10.8	12.7	—	—	—	14.1	—	13.9	19.6	−3	95	7	—	—	—	97	49	—	—
U O 450908	14.1	14.9	14.9	16.3 18.8	—	—	21.5	—	17.1	—	27.6	−1	91	3	—	93	3	—	—	91	67
O E 520434	9.8	9.2	10.2	10.4	—	17.2	14.8	—	—	—	18.9	5	91	9	7	97	7	—	—	—	—
K L 520412	9.7	9.7	10.1	15.0	18.8	18.4	18.6	15.8	—	11.7	19.4	2	82	2	5	90	9	80	46	—	—

*For abbreviations see Table 1, p. 119

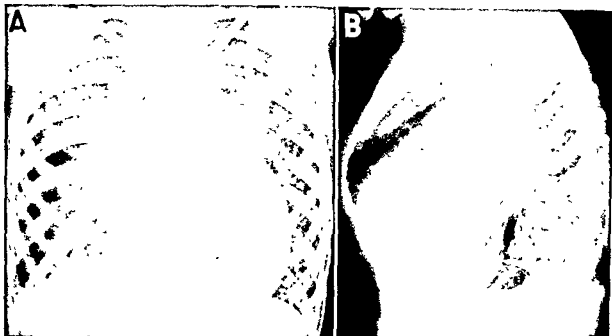


Fig. 703.—Incomplete transposition of the great vessels, ventricular septal defect, levocardia, and transposition of [illegible]
siderable increase in hea[illegible]
the left ventricle is diffi[illegible]
branches in the lungs, normal impression of aortic arch in the esophagus, left-sided superior vena cava; slight enlargement of atria, strongly marked precordial bulge.

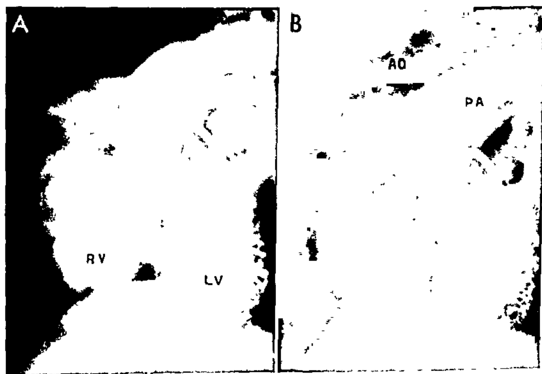


Fig. 704.—Complete transposition of aorta and incomplete transposition of pulmonary ar- [illegible]

tricular septum cannot be determined Angiocardiographic examination with injection of the contrast medium into the right ventricle is therefore required for a more exact anatomic diagnosis In one of our cases the catheter passed only into the aorta, and in two cases only into the pulmonary artery. In the fourth case it entered neither the pulmonary artery nor the aorta, but advanced into the left atrium and ventricle through an atrial septal defect. The left side was also catheterized in another case, in which the catheter passed through a patent foramen ovale. The systolic pressure must be the same in the two ventricles, in the aorta and in the pulmonary artery (Table 30)

The oxygen content usually is higher in samples from the pulmonary artery than from the aorta (292, 502, 672). This applied in one of our two cases in which samples were obtained from both arteries In the other case, however, the oxygen content was the same in samples from pulmonary artery and aorta. The continuous murmur in the latter case may have indicated the presence of a patent ductus, which would have been able to equalize the difference between the oxygen content of the two vessels. The murmur in patent ductus is not, however, usually continuous when the pressure in the pulmonary artery is so high. Nor was it possible to determine by angiocardiography whether or not a patent ductus was present

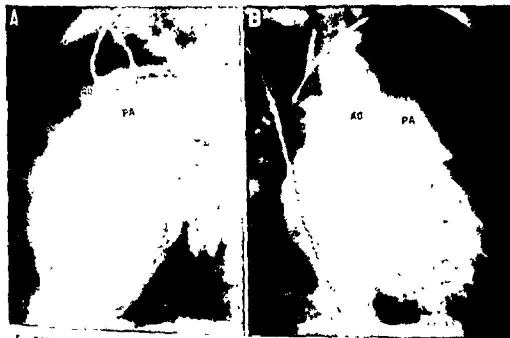


Fig 705.—Incomplete transposition ...

ANGIOCARDIOGRAPHY

The importance of angiocardiography in diagnosis of the Taussig-Bing complex has been stressed by Martin and Lewis (480). In our cases this examination was performed selectively, by injection of the contrast medium into the right ventricle in three cases, and into the left ventricle in the remaining case

The ventricular septal defect was clearly visualized in the lateral view in three of the cases (Fig. 704), and the positional relation of the great vessels to it could be determined. The aorta was given off entirely from the right ventricle, whereas the greatly dilated pulmonary artery had a dorsal origin, although its course was more ventral than is typical in complete transposition (Figs. 704 and 705). The biventricular origin of the pulmonary artery was most apparent in the case illustrated in Figure 704. A similar picture has been published by Gasul *et al.* (270).

In the frontal view, the positional relation of the aorta and pulmonary artery was essentially the same as in ventricular septal defect with a normal origin of the vessels, apart from the high position of the aortic root, due to the transposition.

29 Corrected Transposition of the Great Vessels

IN COMPLETE transposition of the great vessels, the oxygen saturation of the blood in the pulmonary artery is higher than that in the aorta. In functionally corrected transposition there is, on the contrary, a normal route of blood flow through the heart and great vessels, even though—anatomically—the great arteries are transposed. In all cases hitherto described, the aortic orifice has been situated anteriorly to the left and the pulmonary orifice posteriorly to the right, or the aortic orifice anteriorly to the right and the pulmonary orifice posteriorly to the left (359). Correction can take place through *inversion of the bulbar*, ventricular, or sino-atrial region (142).

Ventricular inversion is most common. The pulmonary artery then arises from a ventricle which, *structurally*, has the appearance of a *left* ventricle, but which communicates with *the right* (venous) atrium through an orifice provided with two cusps. The aorta is given off from a ventricle which has the appearance of a right ventricle, but which *receives* blood from the left (arterial) *atrium* through an orifice with three cusps. There *is* inversion of the coronary arteries (538).

As a rule there are coincident malformations, such as valvular *deformities*, a septal defect, or patent ductus *arteriosus* (538, 636, 682). In 1956, Cardell (142) assembled 24 cases from the literature and described one of his own. Corrected transposition without other cardiac malformations was present in only two of 18 cases in which a complete anatomic description was given. In one case there was coincident situs inversus viscerum (538), which, clinically, can arouse false suspicions of mirror-image dextrocardia (257). Additional cases have subsequently been reported by Helmholz *et al.* (329) and Platzer (538). Much has been written about the origin of the malformation (90, 197, 320, 420, 422, 532, 533, 623), but no generally accepted concept exists.

The cause of the peculiar arrangement of the ventricles in corrected transposition is regarded by Lewis and Abbot (431) *to* be that the cardiac tube has bent in the reverse direction to the normal one, so that the aortic limb turns upward on the left side of the common ventricle, instead of the right (local mirror image). Lochte (442), on the other hand, *expressed the* view that it is due to left-hand twisting of the early ventricular loop. Shaner (599) partly adhered to the latter view *after* thorough studies of different types of transposition in the embryonic heart of the pig and comparison of his findings with corrected transposition in an infant who died of bronchopneumonia and meningitis at 10 months of age.

Shaner has stated the cause of transposition of the great vessels to be a delayed descent of the aorticopulmonary (*conus*)

septum. In his opinion, this septum normally grows down in the frontal plane from the region of the fourth to sixth branchial arch arteries toward the conus cushions (1 and 3) and fuses with them. This development is followed by a 90° rotation of the conus cushions and the ventricular loop, the conus septum then being twisted spirally. If growth of the conus septum is somewhat delayed for one reason or another, so that the 90° rotation occurs before the septum has reached the conus cushions, it will instead fuse with the two conus cushions (2 and 4) situated at right angles to the aforementioned ones (1 and 3). In this event, spiral twisting of the septum cannot take place, but the septum remains in the frontal plane. The part of the truncus in front of the septum (aorta) will therefore arise from the right ventricle, and the part behind it (pulmonary artery), from the left ventricle (Fig. 706).

According to Shaner, another type of complete transposition exists, although it is rare. In this type, the great arteries are attached to their usual ventricles, but the ventricles themselves have been reversed by a left-hand twisting of the early ventricular loop. This reverse twisting must occur early, long before division of the primitive atrioventricular canal When this has taken place by fusion of the dorsal and ventral atrioventricular cushions, the right atrium will communicate with the left ventricle (and aorta) and the left atrium with the right ventricle (and pulmonary artery). See Figure 707

Shaner regards corrected transposition as a combination of the two forms just described, a left-hand twist of the ventricular loop and a delayed descent of the aortico-pulmonary septum.

According to Helmholz *et al* (329), coincident mitral incompetence is present in many cases of corrected transposition This is probably related to the left-hand twist of the ventricular loop, and is due to a developmental disturbance of the left atrioventricular valve.

Most of the cases of corrected transposition in the literature have only been diagnosed post mortem, but in recent years detailed descriptions of the clinical features have been given (13, 329). If this malformation is borne in mind, it can easily be diagnosed, particularly with the help of angiocardiography.

The first edition of this book contained two cases of a condition described as a rotation anomaly, combined in one case with valvular pulmonary stenosis and ventricular septal defect, and in the other with mitral incompetence. They were accounted for in the chapters on pulmonary stenosis and mitral incompetence, respectively. With consideration to the findings at examination and the views now prevailing, these cases should, however, rightly be assigned to the chapter on corrected transposition of the great vessels, as also pointed out by Anderson *et al.* (13). An additional two cases have been observed since 1955 All four patients had associated malformations, i.e., (1) mitral incompetence (patient M.N.); (2) pulmonary stenosis and ventricular septal defect (patient B.H.), (3) a large atrial septal defect involving practically the whole septum (patient A.-M E.); and (4) ventricular septal defect and stenosis of the valve between left atrium and right ventricle (patient U.S). In the first patient the picture was so clearly dominated by mitral incompetence that we considered this to justify an account of the patient in Chapter 27. The salient features in the other cases were as follows.

Boy, aged 8½ years (B H. 450218).—Heart disease was diagnosed on a routine examination when the patient was 8 months of age Development was normal He was only slightly disabled, he could play with healthy children of the same age, but his physical activity was somewhat less than theirs. He exhibited slight generalized cyanosis and clubbing of the fingers and toes The physical findings were characterized by bradycardia, a bruit de canon, an inaudible second sound, and a long, high-frequency, loud systolic murmur over the second right intercostal space

Girl, aged 8 years (A-M E 48303) —A murmur was heard at birth, and congenital heart disease was diagnosed Development was normal, and she had never had any cardiac symptoms The physical findings were



Fig. 706 —Uncorrected transposition of the great vessels. Boy aged 3 (H.F. 510828). The aorta (AO) is given off entirely from the right ventricle (RV). Only the part of the vessel close to the valve has an abnormal position. It lies in front of the pulmonary artery (PA) and to the right of it. The ventricle lying anteriorly from which the aorta is given off, has the appearance typical of a right ventricle (D). Slight leakage of contrast medium to left side.

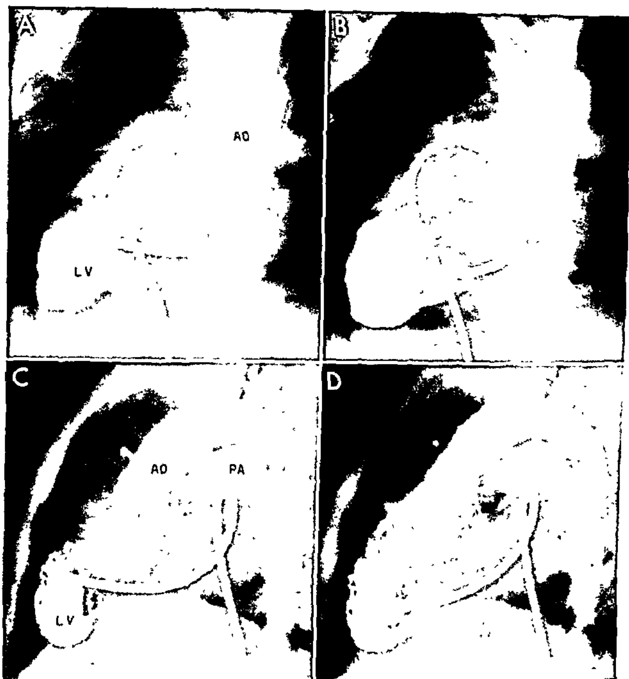


Fig. 707.—Transposition of the ventricles Boy, aged 1 (T N 551105) The catheter advances from the right atrium directly into a chamber (*LV*) with the appearance of a left ventricle, which lies anteriorly to the right, owing to bending of the primitive heart tube and rotation to left instead of to right From this ventricle is given off the aorta (*AO*), which lies in front of and to left of pulmonary artery (*PA*) in position typical of left-hand rotation of the heart tube. Slightly above and to right of *LV* in *A* and *C*, filling defect is visible, it is caused by the tricuspid valve and inflowing blood from the right atrium. Ventricular septum intact Pulmonary artery opacified through patent ductus. (Courtesy of Dr. F Ulfsparre, Kronprinsessan Lovisas Barnsjukhus, Stockholm.)

characterized by a parasternal lift, an accentuated, pure second sound, and a grade 2, early-systolic murmur over the third right intercostal space, as well as a faint diastolic murmur over the fourth right intercostal space

Boy, aged 6 months (U S 550209) —A murmur was heard at 2 months of age, and congenital heart disease was diagnosed Cyanosis and dyspnea were observed from 4 months of age. Weight gain was somewhat poor On examination he exhibited slight generalized cyanosis, severe dyspnea at rest, but *no edema* A slight precordial bulge was present, a pure second sound, more marked over the "pulmonary area" than over the "aortic area," and a grade 5, systolic and diastolic (to and fro) murmur over the fourth left intercostal space. He died a few days after the investigations were completed The autopsy findings can be inferred from Figure 710b

The pulmonary orifice lies more dorsally and caudally and slightly more to the right than is normal. If the pressure in the pulmonary artery is normal, the pulmonary component of the second sound is inaudible. The aortic component, on the contrary, is heard as an accentuated, pure second sound over the "pulmonary area." To the right of the sternum, the second sound is faint. This can be inferred from Figure 708, showing the phonocardiogram in the second case, in which there was an atrial septal defect with a large arteriovenous shunt and normal pressure in the pulmonary artery. The systolic murmur was loudest one intercostal space farther down than is customary in atrial septal defect. The intensity of the murmur was low, which is explained by the great distance between the pulmonary artery and the thoracic wall. Its high frequency nevertheless shows that the velocity of flow through the pulmonary orifice was high, i.e, the stroke volume was large

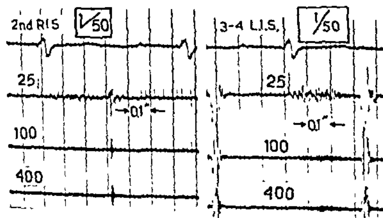


Fig 708 —Phonocardiogram in corrected transposition of the great vessels ... defect Girl ...
... space, R I S , right interspace

CLINICAL FEATURES

Physical Signs

Although in these cases the physical findings are chiefly characteristic of the associated malformations, certain of them are attributable to the transposition

The aortic orifice lies more ventrally, cranially, and to the left than normal The position corresponds approximately to the normal position of the pul...

Electrocardiography

An A-V block has been found in many of

According to Anderson *et al.* (13), a reversal of the QRS pattern, with QR in lead V_1 and RS in lead V_6, is another typical change. High voltage of the P waves in lead II was also a fairly constant feature in their series.

In our small series, complete atrioventricular dissociation was present in one case (B.H.) and a first degree A-V block with a P-R interval of 0 22 sec in two cases (M.N. and A-M.E.). The P-R interval was normal in only one case (U.S.). This is apparent from Figure 709, which shows the *electrocardiograms in all the cases,* including that with associated mitral incompetence.

The electrocardiographic pattern is dependent partly on the hypertrophy of the ventricles that may be caused by associated anomalies and partly on the position of the various chambers of the heart in the thorax. Both factors vary appreciably. The rotation of the heart is highly individual (cf. Angiocardiography). If no associated malformation is present, the left ventricle works under an abnormally low pressure

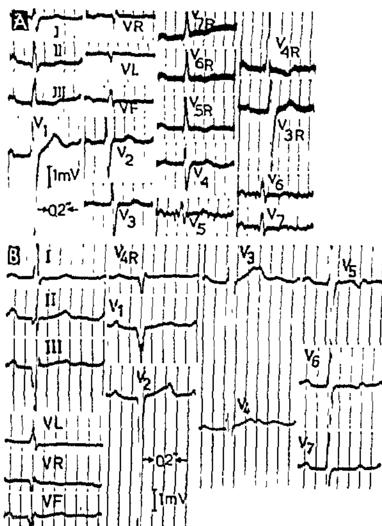


Fig. 709.—Electrocardiograms in corrected transposition of the great vessels *A*, girl, aged 8 (A-M E. 480303), with associated atrial septal defect and normal pulmonary artery pressure *B*, girl, aged 8 (M N. 450424), with associated regurgitation in valve between left atrium and right ventricle The pulmonary artery pressure was normal C, boy, aged 8½ (B H 450218), with associated pulmonary stenosis and ventricular septal defect *D*, boy, aged 6 months (*U S* 550209), *with associated ventricular septal defect and stenosis of the valve between left atrium* and right ventricle (*continued*)

(pulmonary circuit) and with a normal stroke volume, whereas the right ventricle works under a pressure that is abnormally high for this chamber (systemic circuit).

In two of our cases the pulmonary artery pressure was normal, in one (A-M.E) there was an atrial septal defect, and in the other (M.N., see Chap. 27), incompetence of the valve between the left atrium and right ventricle. Thus the stroke volume tiguous to the anterior thoracic wall (see Angiocardiography, Fig. 715). Figure 709, *A* shows the electrocardiogram in this case. The transitional zone lies between leads V_{7R} and V_1. In leads V_{5-7} the QRS complex has the appearance that can be seen over a hypertrophied right ventricle, and in leads V_{3R-7R} the QRS pattern can be explained by dilatation of the left ventricle.

In the other case (M.N.) both ventricles

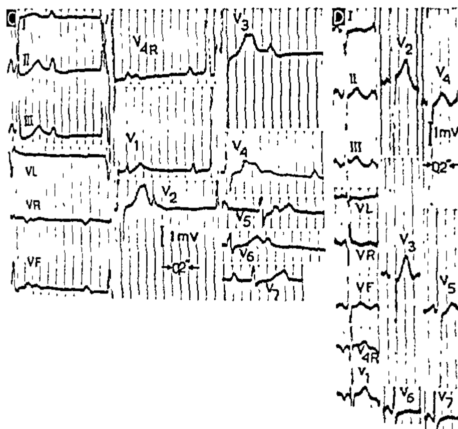


Fig. 709 *(cont.)*

of the left ventricle was increased in the former case, and that of the right ventricle, in the latter. The position of the ventricles was entirely different in the two cases. In the former the ventricular septum lay somewhat to the right of the midline, approximately in the sagittal plane, with the left ventricle entirely to the right of the midline and the right ventricle mainly to the left of it. Both ventricles were con- lay entirely to the left of the midline, the left to the right and slightly in front of the right ventricle, and the septum at an angle of about 45° to the sagittal plane. The orifice between right atrium and left ventricle lay in the sagittal plane, directly to the left of the midline. Consequently, ventricular cavity complexes were obtained in leads V_{4R} and V_1 (Fig. 709, *B*). Only a low voltage of R was recorded over the thin-walled

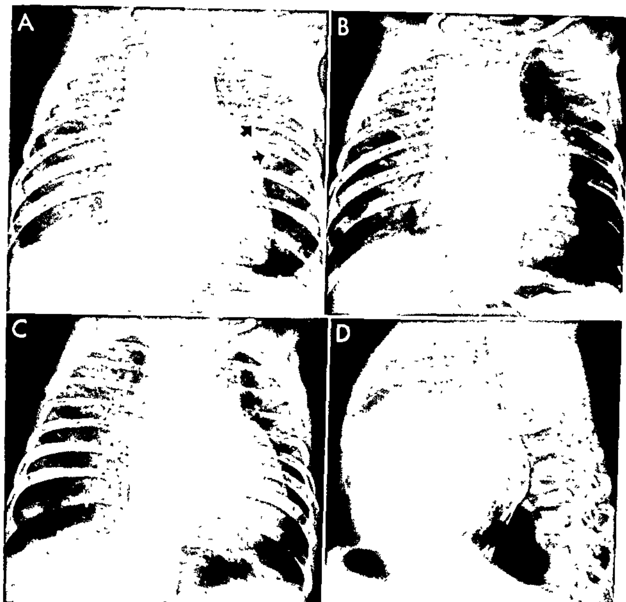


Fig. 710a.—Corrected transposition of the great vessels and ventricular septal defect Boy, aged 6 months (U S 550209) see Figure 710b The large left atrium produces a local bulge in the left outline (arrows in *A*). Hypertrophy of right ventricle with forward bulging of anterior thoracic wall. Increased blood volume in pulmonary circulation.

left ventricle. Tall R spikes with S-T and T deviation were, on the contrary, recorded over the hypertrophied, dilated right ventricle in leads V_{6-7}.

A large ventricular septal defect was present in two cases. In one (B.H) it was combined with pulmonary stenosis and in the other (U.S.) with stenosis of the atrioventricular valve between left atrium and right ventricle. In the former case the position of the ventricles was the reverse of the normal one, and the ECG showed reversal of the QRS pattern and signs of right ventricular hypertrophy (Fig 709, *C*). In the latter case the positional relation of the ventricles was fairly normal, and the ECG showed the features of right ventricular hypertrophy and an incomplete right bundle-branch block (Fig. 709, *D*).

ROENTGENOLOGIC EXAMINATION

As pointed out by Anderson *et al* (13), the size and shape of the heart in corrected

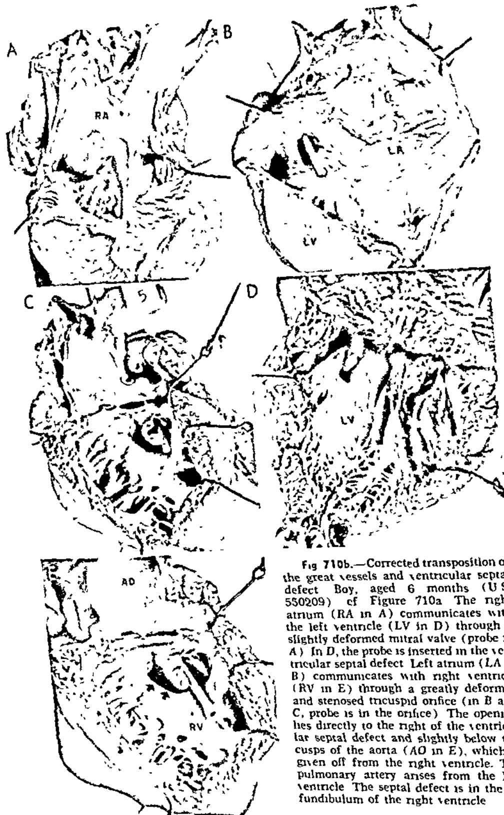


Fig 710b.—Corrected transposition of the great vessels and ventricular septal defect Boy, aged 6 months (U S 550209) cf Figure 710a The right atrium (RA in A) communicates with the left ventricle (LV in D) through a slightly deformed mitral valve (probe in A) In D, the probe is inserted in the ventricular septal defect Left atrium (LA in B) communicates with right ventricle (RV in E) through a greatly deformed and stenosed tricuspid orifice (in B and C, probe is in the orifice) The opening lies directly to the right of the ventricular septal defect and slightly below the cusps of the aorta (AO in E), which is given off from the right ventricle. The pulmonary artery arises from the left ventricle The septal defect is in the infundibulum of the right ventricle

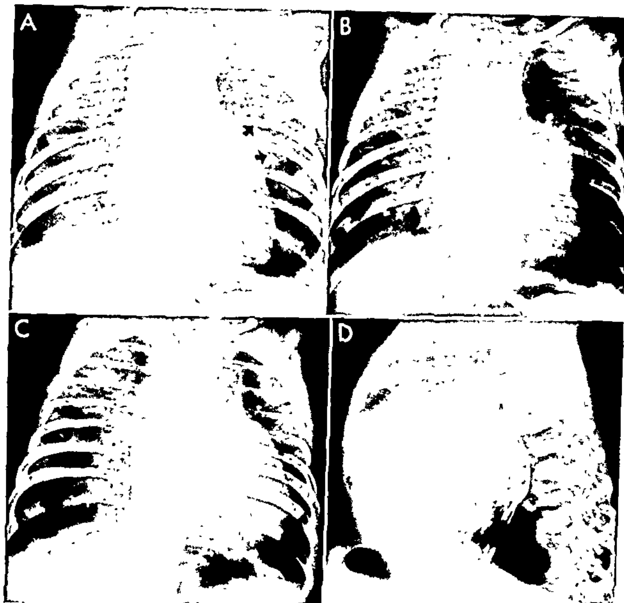


Fig. 710a.—Corrected transposition of the great vessels and ventricular septal defect. Boy, aged 6 months (U S 550209): see Figure 710b The large left atrium produces a local bulge in the left outline (arrows in *A*) Hypertrophy of right ventricle with forward bulging of anterior thoracic wall. Increased blood volume in pulmonary circulation

left ventricle. Tall R spikes with S-T and T deviation were, on the contrary, recorded over the hypertrophied, dilated right ventricle in leads V_{6-7}.

A large *ventricular septal defect* was present in two cases. In one (B H) it was combined with pulmonary stenosis and in the other (U S.) with stenosis of the atrioventricular valve between left atrium and right ventricle. In the former case the position of the ventricles was the reverse of the normal one, and the ECG showed reversal of the QRS pattern and signs of right ventricular hypertrophy (Fig. 709, *C*). In the latter case the positional relation of the ventricles was fairly normal, and the ECG showed the features of right ventricular hypertrophy and an incomplete right bundle-branch block (Fig. 709, *D*)

ROENTGENOLOGIC EXAMINATION

As pointed out by Anderson *et al.* (13), *the size and shape of the heart in corrected*



Fig 710b —Corrected transposition of the great vessels and ventricular septal defect Boy, aged 6 months (U S 550209) cf Figure 710a The right atrium (*RA* in *A*) communicates with the left ventricle (*LV* in D) through a slightly deformed mitral valve (probe in A) In D, the probe is inserted in the ventricular septal defect Left atrium (*LA* in B) communicates with right ventricle (*RV* in *E*) through a greatly deformed and stenosed tricuspid orifice (in *B* and C, probe is in the orifice) The opening lies directly to the right of the ventricular septal defect and slightly below the cusps of the aorta (AO in E), which is given off from the right ventricle The pulmonary artery arises from the left ventricle The septal defect is in the infundibulum of the right ventricle

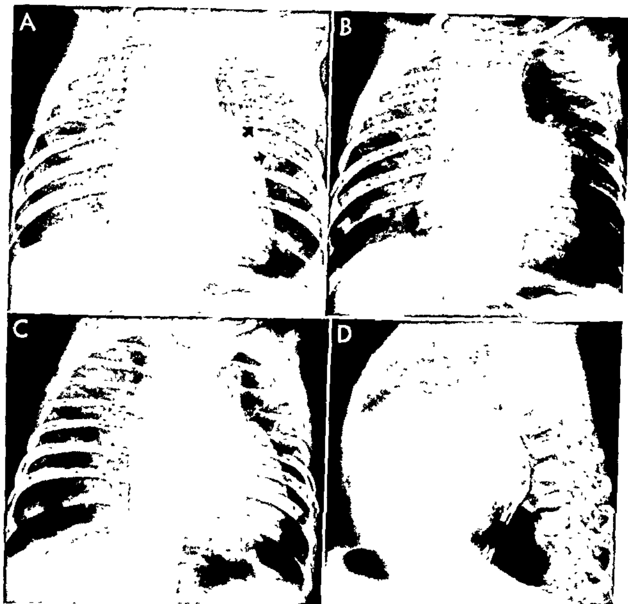


Fig 710a.—Corrected transposition of the great vessels and ventricular septal defect Boy, aged 6 months (U.S 550209) see Figure 710b The large left atrium produces a local bulge in the left outline (arrows in *A*) Hypertrophy of right ventricle with forward bulging of anterior thoracic wall Increased blood volume in pulmonary circulation

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ROENTGENOLOGIC EXAMINATION

As pointed out by Anderson *et al.* (13), the size and shape of the heart in corrected

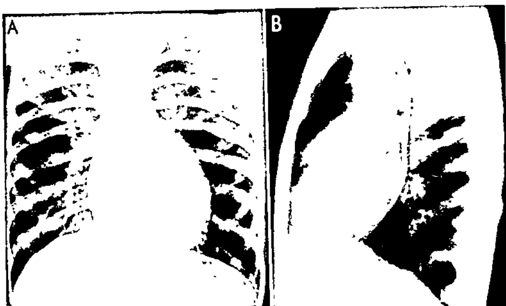


Fig 712 —C
stenosis, and c
heart is rotated
aorta (cf Fig

Fig 713 —Corrected transposition of the great vessels A, boy, aged 8
aged 8 (M N 450424)
sion in the esop
in B) Upper arr

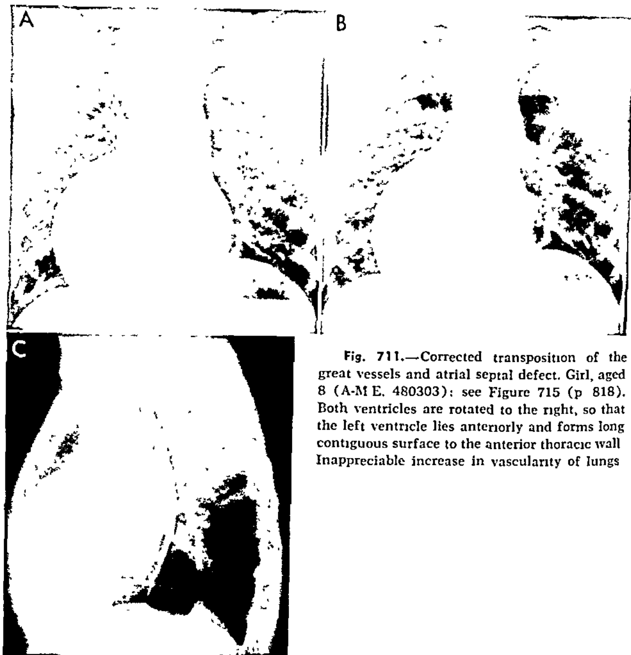


Fig. 711.—Corrected transposition of the great vessels and atrial septal defect. Girl, aged 8 (A-M E. 480303): see Figure 715 (p 818). Both ventricles are rotated to the right, so that the left ventricle lies anteriorly and forms long contiguous surface to the anterior thoracic wall Inappreciable increase in vascularity of lungs

transposition of the great vessels are largely determined by the associated malformations. Consequently, no uniform, characteristic picture exists, but the picture varies from case to case (Figs. 677a and 710–712). However, when those features that are directly due to the abnormal position of the great vessels and ventricles are conspicuous, there are good reasons for making a presumptive diagnosis of corrected transposition. Thus the cardiac apex is often broad and upturned (Figs 677a and 712). The left superior cardiovascular outline, which in this condition is not infrequently formed by the aorta alone, is strikingly elongated and has a slightly convex (Fig 712) or faintly concave course (Fig. 677a). The pulmonary artery generally cannot be identified as a separate outline. This finding seems to apply even if the artery is poststenotically dilated Owing to its medial position, it often produces an impression in the esophagus from the left (Fig. 713) or displaces a long segment of

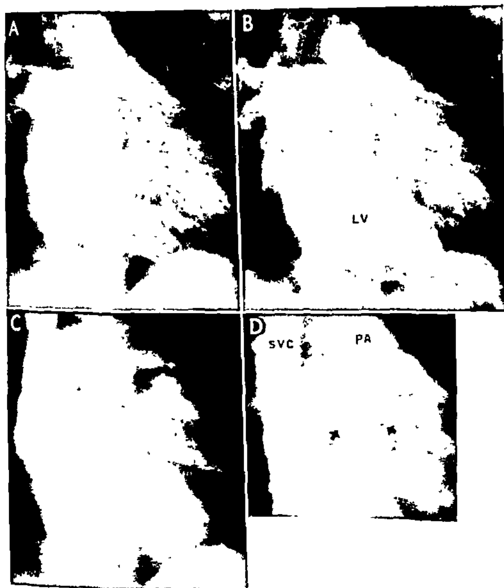


Fig 714 —Corrected transposition of the great vessels, ventricular septal defect, pulmonary stenosis, and complete atrioventricular block ...
tion of the ventricles, s
with both right atrium
tricular septal defect ...
and is given off fror
in D point to valv
(*continued*)

this structure. The impression made by the aortic arch in the esophagus is ordinary or may be lacking (Fig. 713).

With coincident stenosis of the arterial arterioventricular orifice, the picture is, however, dominated by the enlargement of the left atrium (Fig. 710), and the aforementioned features are not apparent.

CARDIAC CATHETERIZATION

The abnormal position of the orifices, great vessels, and ventricles can sometimes be established merely on the basis of the passage of the catheter (329). The position of the catheter must, however, be studied in both the frontal and lateral views. The situation of the semilunar orifices is the most typical and constant feature (cf Angiocardiography), whereas that of the ventricles may vary appreciably. It may nevertheless often be difficult to advance the catheter into the pulmonary artery. If there is a large septal defect with a bidirectional shunt, it may be practically impossible to determine by gas analysis whether the catheter lies in the venous or arterial ventricle In such cases, the angiocardiograms are decisive for the diagnosis.

Catheterization not infrequently provides valuable information about associated malformations Table 31 shows the findings at cardiac catheterization in three of our cases, the fourth case is accounted for in Table 29 (p 769) Only in the last-mentioned case was it possible to advance the catheter into the pulmonary artery. This was one of the earliest cases in our series of cardiac catheterizations We had observed the abnormal position of the catheter, but had interpreted it as due only to a rotation anomaly of the heart.

Patient A-M E. had a defect involving practically the whole atrial septum There was not only a very large left to right shunt, but also a small right to left shunt, with an arterial oxygen saturation of 92 per cent, as compared with 96 per cent in the pulmonary venous blood The pressure in the venous ventricle was not raised

In two patients the pressure in this ven-

TABLE 31 —CORRECTED TRANSPOSITION OF THE GREAT VESSELS: FINDINGS ON CARDIAC CATHETERIZATION IN 3 CASES*

Case	O_2 Content, vol %							Art O_2 Sat, %	O_2 Cap, vol %	Pressure, mm Hg							
	SVC	IVC	RA	LA	PV	Ventricles				RA	LA	Ventricles				Aorta	
						Venous	Arterial			Mean	Mean	Venous Syst	Venous End-diast	Arterial Syst	Arterial End-diast	Syst.	Diast
A-M E 480303	14 4	13 4	16 0	18 3	18.3	16 0	17 5	92	18 8	5	5	31	2	86	6	—	—
L-B H 450218	11.7	13 4	12 3	20 4	21 0	12 8	20 3	81	21.9	7	9	100	6	108	6	—	—
U S 550209	6.7	7 0	6 7	—	—	7.8	—	79	13 5	3	—	65	4	—	—	60	42

*For abbreviations see Table I, p 119

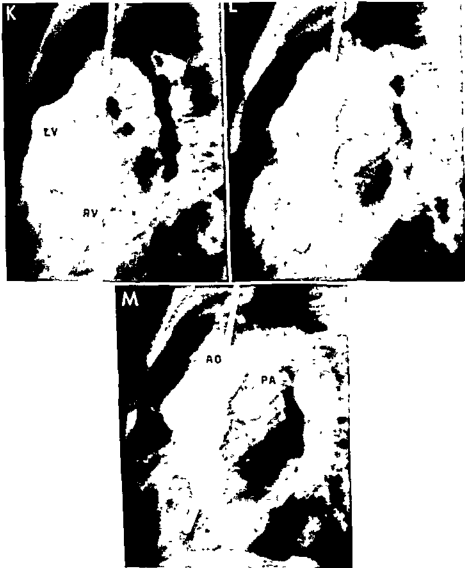


Fig. 714 (cont)

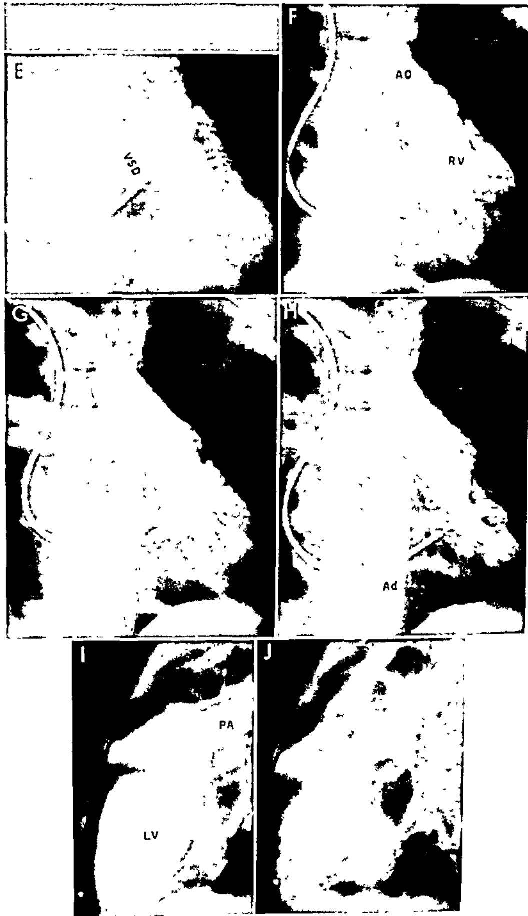


Fig. 714 (*cont.*)



Fig. 715 (*cont.*)

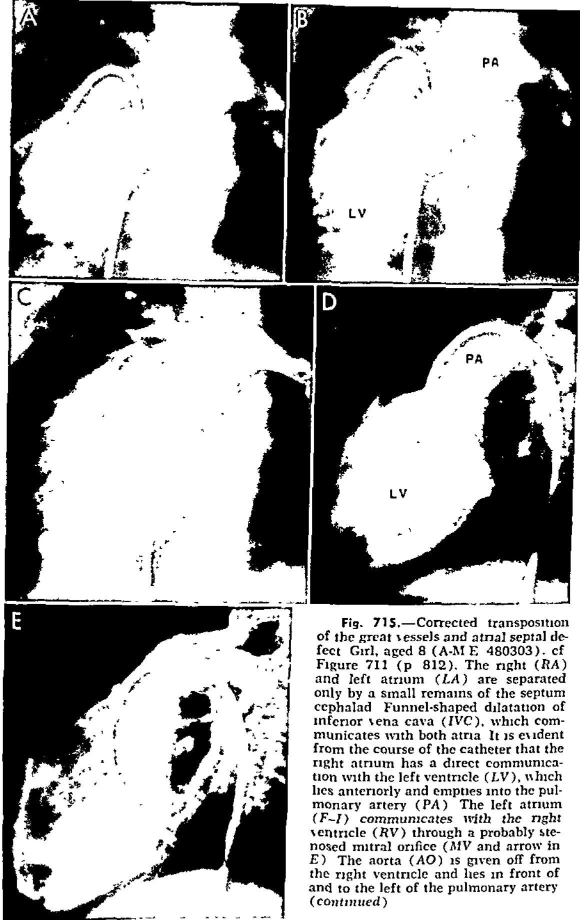


Fig. 715.—Corrected transposition of the great vessels and atrial septal defect Girl, aged 8 (A-M E 480303). cf Figure 711 (p 812). The right (*RA*) and left atrium (*LA*) are separated only by a small remains of the septum cephalad Funnel-shaped dilatation of inferior vena cava (*IVC*), which communicates with both atria It is evident from the course of the catheter that the right atrium has a direct communication with the left ventricle (*LV*), which lies anteriorly and empties into the pulmonary artery (*PA*) The left atrium (*F–I*) *communicates with the right* ventricle (*RV*) through a probably stenosed mitral orifice (*MV* and arrow in *E*) The aorta (*AO*) is given off from the right ventricle and lies in front of and to the left of the pulmonary artery (*continued*)

30 Primary Pulmonary Hypertension

RAISED PRESSURE in the pulmonary artery may be found in congenital heart disease with a left to right shunt, left ventricular failure, mitral stenosis, diffuse embolism of the pulmonary artery, schistosomiasis, periarteritis nodosa, and in chronic pulmonary disease Pulmonary hypertension may also exist in the absence of any primary disease of the heart or lungs It is caused by narrowing of the small arteries in the pulmonary circulation In order to maintain an adequate cardiac output, the pressure must rise. This disease, which is generally denoted as primary pulmonary hypertension, is somewhat rare. Each author has reported only a few cases (56, 92, 151, 172, 208, 209, 245, 285, 304, 324, 404, 602, 696, 720)

Anatomically, there are changes in the small arteries of the type described in congenital heart disease with a left to right shunt (see p 339) The lumen is greatly constricted, owing to intimal and medial thickening Moreover, the vessels are often thrombosed, giving rise to severe obstruction in the pulmonary circulation The large arteries, on the contrary, are not involved

The pathogenesis is unknown In the discussion of high pulmonary vascular resistance [illegible]
it has be[illegible]
age pre[illegible]
large bl[illegible]
[illegible] is because in some cases it is present from the first months of life, whereas in others increased resistance has not developed even in adult age, despite a large blood flow. Another factor, possibly congenital, seems to be responsible. Primary pulmonary hypertension may occur even in infancy (56), as in one of our cases.

CLINICAL FEATURES

The signs and symptoms are caused by the difficulty in maintaining an adequate cardiac output and by the elevated pressure in the pulmonary circulation. They often appear only on exertion and are chiefly in the form of increased fatigability, syncope, angina pectoris, dyspnea, tachycardia, hemoptysis, and cyanosis.

Right ventricular hypertrophy develops secondarily to the pulmonary hypertension, and congestive failure gradually supervenes Death may be sudden.

In *our series*, three cases of primary pulmonary hypertension were diagnosed One patient (M J 490620), reported on in the previous edition, exhibited deviating features There were localized constrictions of the arterial branches in the periphery of the hilum, with poststenotic dilatation. A faint continuous murmur was audible round the whole thorax, an auscultatory finding that has earlier been described in a case with peripheral stenosis of the pulmonary artery (490). This case has there-

tricle was, on the contrary, as high as in the systemic circulation. Both patients had a large ventricular septal defect. In one of them (B.H.) it was combined with valvular pulmonary stenosis and a veno-arterial shunt, and in the other (U.S.), with stenosis of the atrioventricular valve, between the left atrium and the arterial (right) ventricle, and a mixed interventricular shunt. Stenosis of the atrioventricular valve probably reduced the flow from left atrium to right ventricle, which favored the occurrence in diastole of a veno-arterial shunt from left to right ventricle through the ventricular septal defect. In systole, on the other hand, the shunt presumably passed in the opposite direction The patient had both a systolic and a diastolic murmur, of the nature of a to and fro murmur. The diastolic component differed altogether from the diastolic murmur characteristic of mitral stenosis. The to and fro murmur can possibly be explained by the shunt in both systole and diastole. At autopsy, all the semilunar valves were found to have a normal appearance.

ANGIOCARDIOGRAPHY

Angiocardiography is the simplest and most certain clinical method for diagnosis of corrected transposition of the great vessels. In view of the variable position of the chambers of the heart, we—like Anderson *et al* (13)—have found that the best information generally is provided by exposures in the true frontal and lateral views The abnormal position of the ventricles makes it, however, difficult to determine their identity. We have used the following as criteria of a right ventricle a distinctly developed infundibulum, a crista supraventricularis bulging into the lumen, and in the sinus region, reticular trabeculation more or less clearly related to the papillary muscles The criteria of the left ventricle were absence of an infundibulum or only a very short one, no crista supraventricularis, more longitudinal and less marked trabeculation, and a more triangular shape with the base facing cephalad (Fig. 714).

Angiocardiographic examination was performed in three of our cases. In all of them the pulmonary artery lay behind the aorta and to the right of it (Figs. 677b, 714, and 715). The pulmonary artery arose from the superior part of the left ventricle, fairly far dorsally (Figs. 677b, 714, and 715). The aorta, on the other hand, was given off from the right ventricle, to the left of the pulmonary artery and in front of it. The ascending aorta ran obliquely upward and medially toward the aortic arch, which was at the usual site (Figs. 677b and 714).

In one case (A-M.E.) there were also an extremely large atrial septal defect and a greatly dilated inferior vena cava opening into both right and left atrium (Fig 715). The arterial atrioventricular orifice was stenosed and converted into an elongated funnel

In the second case (B H) a large ventricular septal defect was present (Fig 714). On injection into the right ventricle, a large proportion of the contrast medium passed into the left ventricle, instead of opacifying the right. On injection into the left ventricle, this chamber was completely opacified, with only inappreciable leakage into the right ventricle, probably owing to the difference between the diastolic pressure in the pulmonary artery and that in the aorta. This difference applies irrespective of whether or not an associated ventricular septal defect is present in transposition of the great vessels

In the third case (M N) there was associated incompetence of the arterial atrioventricular orifice (mitral incompetence) During ventricular systole, marked regurgitation of contrast medium took place from the right ventricle into the enlarged left atrium (cf. C, systole, and *D-E*, diastole, in Fig. 677b).

had very pronounced hypertrophy, whereas only slight changes were present in the patient with the lowest pressure in the pulmonary artery (B A. 370730) An abnormally tall P wave in V_1 was present in one case. In two there were also indications of an incomplete right bundle-branch block.

ROENTGENOLOGIC EXAMINATION

The roentgenologic appearance is characterized by enlargement and hypertrophy order—were, on the contrary, of normal width and exhibited normal pulsations. The peripheral vessels of the lungs were very narrow, and an abrupt diminution in caliber in the periphery of the hilum was the most striking roentgenologic feature. These changes in the vascular anatomy were equally marked in all parts of the lungs and were apparently of a similar degree in all the cases

There was considerable curvature of the anterior surface of the right ventricle, as



Fig 717.—Primary pulmonary hypertension. Boy, aged 11 (L O 430514), see Figure 725. Hypertrophy of right ventricle, considerable dilatation of main trunk of pulmonary artery. Appearance of the central branches is normal, whereas peripheral vessels of the lungs are extremely narrow Aorta of normal width, no enlargement of heart

of the right ventricle, dilatation of the pulmonary artery, and narrowness of the peripheral vessels of the lungs In advanced cases, the right atrium is dilated The aorta is usually narrow As a rule, the pulsations in the main trunk of the pulmonary artery are large

In our cases, the dilatation of the pulmonary artery was pronounced and the pulsations extremely large, except in the oldest patient The central branches—corresponding to the branches of the secondary seen in hypertrophy In every case there was an increase in the contiguity of the heart to the anterior wall of the thorax, as an expression of the ventricular enlargement (Figs 717–719).

Possible slight enlargement of the right atrium was seen in one case; the appendage

d in-

two

cases and of normal width in the remaining case The heart volume was moder-

fore been classified as stenosis of the pulmonary artery branches.

GIRL, AGED 18 YEARS (E S. 330606).—She was asymptomatic until 10 months of age, when she contracted severe pertussis complicated by pneumonia. After this illness, a marked exacerbation took place. She was easily fatigued, was unable to sit up, and suffered from tachycardia. Her physical development was retarded, and she was never able to play like a healthy child. A murmur was noted at 7 years of age. During the past few years, her condition had become steadily worse, she was so severely disabled that she could scarcely walk up one flight of stairs On several occasions she had typical attacks of angina pectoris Since 3 years of age she had exhibited cyanosis on exertion, during the past year it was also present at rest Apart from pertussis and pneumonia at 10 months of age, she had had no pulmonary illness

GIRL, AGED 16 YEARS (B A 370730) —She had been apparently healthy For the past two years she had suffered from dyspnea and tachycardia on exertion and could manage only three flights of stairs

BOY, AGED 11 YEARS (L O 430514).—Slight disability had been present since he was 2 to 3 years old, he could run only about 20 meters and this always produced fatigue and dyspnea He had never been cyanotic

In none of the cases was a primary pulmonary disease found on clinical or roentgenologic examination. In case E S. 330606, spirometry showed slightly raised functional residual capacity. The oxygen consumption per liter of ventilation was low at rest, but particularly at work; this is in agreement with the findings of Dresdale *et al.* (208).

PHYSICAL SIGNS

The most characteristic feature is an accentuated second sound over the pulmonary area. It may be so marked that it is easily palpated over the second left intercostal space. The intensity of the second sound depends not only on the pressure in the pulmonary artery, but on the distance between the valves and the stethoscope. Right ventricular hypertrophy is manifested as a parasternal lift of varying degree.

The murmurs are not typical and may be lacking entirely, as was noted in one of our cases. Two of them presented a faint, systolic murmur over the pulmonary area and a faint protodiastolic murmur as well An atrial sound gallop was heard in the clinically most severe case (Fig. 716). An early systolic click over the pulmonary area, found in many cases of pulmonary hypertension associated with a left to right shunt, was not audible in any of these cases of primary pulmonary hypertension.

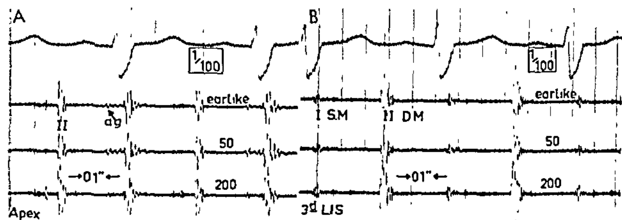


Fig. 716.—Phonocardiograms in primary pulmonary hypertension. Girl, aged 18 (E S 330606) *A*, apex Very faint late systolic murmur, atrial sound gallop (*ag*) *B*, 3rd left interspace (*3rd L I S.*). *Highly accentuated second sound (II), faint protodiastolic murmur (DM)* Boxed figures denote degree of amplification, other figures denote standard frequencies of filters

ELECTROCARDIOGRAPHY

The ECG is characterized by right ventricular hypertrophy. Two of our patients

ately increased in two cases and normal in the third.

Roentgenologic findings mainly in agreement with the observations in our cases have been reported by Dresdale *et al.* (208, 209) They pointed out that the central vessels of the lungs may also be dilated, a feature which seems to be more common in adults than in children. As a rule the peripheral pulmonary vessels are narrow, but it has been stated that they may have a normal appearance. We are, however, inclined to believe that the change in the caliber of the vessels in the periphery of aortic root. Moreover, in both conditions there are generally large pulsations in the pulmonary artery. It is possible, however, to differentiate between pulmonary hypertension and pulmonary stenosis by means of electrokymographic recordings of the pulsations in the pulmonary artery. Presumably, this differential diagnosis is easy from the clinical point of view.

ELECTROKYMOGRAPHY

Characteristic electrokymograms from the pulmonary artery can be recorded in



Fig 720.—Electrokymograms in primary pulmonary hypertension Boy, aged 11 (L O 430514) PCG over pulmonary area. *Left*, the pulmonary artery curve is characterized by a slow

pearance. I, 1st sound, *II*, 2nd sound

the hilum has not been taken sufficiently into account in the evaluation of these cases

The roentgenologic appearance in primary pulmonary hypertension is characteristic but not specific Exactly the same features may be found in other forms of pulmonary hypertension, e g , in patent ductus arteriosus, in atrial septal defect, and in ventricular septal defect, particularly in cases in which the shunt is entirely or dominantly reversed An identical roentgenologic appearance and the same fluoroscopic findings may also be present in valvular pulmonary stenosis with normal many forms of pulmonary hypertension. In our three cases of primary pulmonary hypertension so studied, such typical tracings were also observed

The common features of the curves were: a slow rise of the systolic limb, with the summit at the end of systole; a small dicrotic wave; and an abnormal position of the incisura, high up on the diastolic limb, in the immediate vicinity of the crest. These also characterize the pressure curves from the pulmonary artery. The dicrotic wave on the electrokymogram was greatly diminished, and a distinct incisura was recorded in only one case (Fig 721) In the two



Fig. 718.—Primary pulmonary hypertension. Girl, aged 16 (B A. 370730). Hypertroph of right ventricle, dilatation of main trunk of pulmonary artery, central vessels of normal widtl peripheral vessels very narrow, narrow aorta, slight increase in heart volume

A B

Fig. 719.—Primary pulmonary hypertension Girl, aged 18 (E S 330606) Great hypertrophy of right ventricle and marked curvature of anterior surface of heart, considerable dilatation of main trunk of pulmonary artery Width of the central vessels is unchanged, whereas vessels in the periphery are extremely narrow Aorta narrow, slight enlargement of right atrium, appendage prominent, moderate increase in heart volume

temic pressure argues in favor of a ventricular septal defect, but does not exclude primary pulmonary hypertension. In cases with practically equilibrated pressure in the ventricles a ventricular septal defect can always be demonstrated on angiocardiography by *rapid* injection directly into the right ventricle, even if there is no right to left shunt. Since the injection also takes place during diastole, the contrast medium is forced over to the left ventricle.

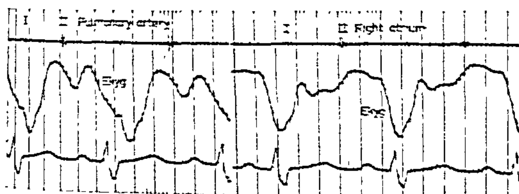


Fig 722.—Electrokymograms in primary pulmonary hypertension. Girl, aged 18 (E.S. 330606). PCG over pulmonary area. *Left*, the pulmonary artery curve shows a somewhat slow upstroke, with crest of the curve closer to the end of systole than normally. The incisura and dicrotic wave cannot be identified. The positive diastolic wave does not correspond to a dicrotic wave, since it occurs too late in relation to the 2nd pulmonary [illegible] (damped curves [illegible] corresponds to a [illegible] corded, but in vi[illegible] The curve is inc[illegible] the right ventric[illegible] 2nd sound.

TABLE 32.—PRIMARY PULMONARY HYPERTENSION: MOST IMPORTANT FINDINGS ON CARDIAC CATHETERIZATION IN 3 CASES*

Case	Pressure mm Hg							Art O₂ Saturation %
	RV	PA	PCV	LA	LV	PCA	Systemic Artery	
E.S. 330606 (18 yr.)	64	65/53 (damped)	6	—	—	—	130/100†	73
B.A. 370730 (16 yr.)	51/6	53/29	6	5	112/11	16 (damped)	—	96
L.O. 430514 (11 yr.)	103/8	96/46	3	—	—	—	100/53	96

*For abbreviations see Table 1 p. 119
†Measured with a cuff.

Cardiac catheterization was performed in our three cases (Table 32), and a left to right shunt could then be excluded. In one case (E.S. 330606), there was arterial oxygen unsaturation, but no right to left shunt could be demonstrated on venous angiocardiography. Consequently, deficient oxygenation of the blood in the lungs is the only explanation of the anoxia.

In most cases, however, decreased arterial oxygen saturation has been regarded as a result of a right to left shunt through a patent foramen ovale.

In one case the pressure in the pulmonary circulation was on the same level as that in the systemic circulation, which is highly indicative of ventricular septal defect. An intact ventricular septum could, however, be demonstrated on angiocardi-

others, it was, respectively, almost entirely flattened and obliterated (Figs. 720 and 722). In one of these, an extra wave was recorded in diastole; it may have been reflected from the periphery. The onset of upstroke followed the Q wave after a normal interval and was well defined in all the cases.

In one case, the electrokymogram recorded over the right atrium showed alterations corresponding to a changed volume-pressure relationship in the atrium, with increased presystolic activity (Fig 722) In the two others, the appearance of the atrial tracings was normal (Figs 720 and 721).

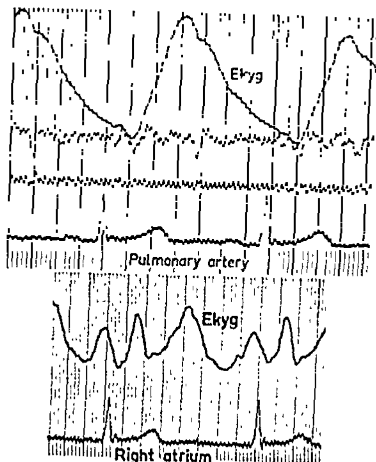


Fig. 721.—Electrokymograms in primary pulmonary hypertension Girl, aged 16 (B A 370730). *Upper tracings*, sinus-shaped pulmonary artery curve The incisura is situated high up. Reduced dicrotic wave. *Lower tracings*, configuration of the right atrial curve is normal

CARDIAC CATHETERIZATION

It was only with the introduction of cardiac catheterization that it became possible to diagnose pulmonary hypertension with certainty and to make an exact estimate of its degree. A diagnosis of primary pulmonary hypertension can, however, be made only after other causes of a rise in pressure in the pulmonary artery have been ruled out. By catheterization it can be established whether a left to right shunt exists or a raised PCV pressure is present as an indication of primary disease of the left side of the heart. But it is not always possible, even on catheterization, to rule out a ventricular septal defect. The resistance in the pulmonary circulation may be so high that no left to right shunt occurs As a rule, a small right to left shunt is present in such cases, but in primary pulmonary hypertension as well the arterial oxygen saturation may be slightly lowered A systolic pressure in the right ventricle on the same level as the sys-

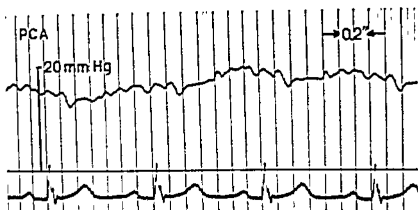


Fig 724 —PCA pressure curve in primary pulmonary hypertension Girl, aged 16 (B.A. 0730) The curve does not have the typical appearance of an arterial pulse curve, but is mped (cf. Fig 413, p. 446).



Fig 725 —Primary pulmonary hypertension Boy, aged 11 (L O. 430514) Hypertrophy of ight ventricle, considerable dilatation of main trunk of pulmonary artery and its main branches, brupt diminution in vessel caliber in periphery of the hilum, peripheral pulmonary vessels extremely narrow

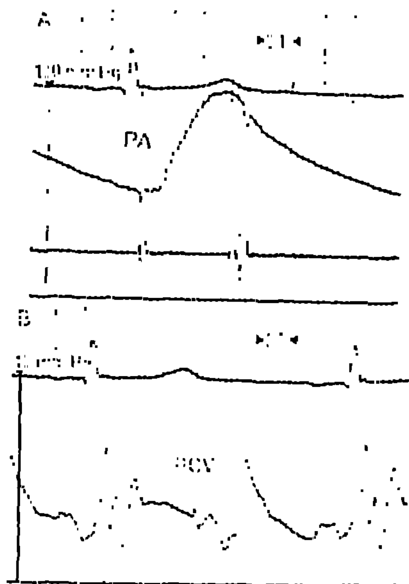


Fig. 723.—Primary pulmonary hypertension Boy, aged 11 (L O 430514) *A*, the pressure curve from the pulmonary artery shows a slow rise, as well as a slow descending limb The incisura lies close to the summit of the curve. *B*, the PCV pressure is not raised. The shape is not that of a venous pulse curve, but exhibits uncharacteristic deflections.

ography with rapid injection into the right ventricle.

When peripheral resistance is high, the pressure curve from the pulmonary artery has a characteristic shape. The pressure rises slowly, so that the summit appears late The incisura lies high on the descending limb, which declines slowly. The curve becomes more sinus-shaped (Fig 723, *A*).

The PCV pressure was not obtained in one case, but there were no signs of a valvular lesion in the left side of the heart, and the left atrium was not found to be enlarged on roentgenologic examination. In the other cases the pressure was not raised, thus showing that the pulmonary hypertension was not due to lesions of the left side of the heart The curve did not have the shape of a typical venous pulse curve, but exhibited uncharacteristic deflections (Fig 723, *B*). The PCA pressure could be recorded in one of our cases (Fig. 724). The curve did not resemble an arterial curve, but was greatly damped. This find-

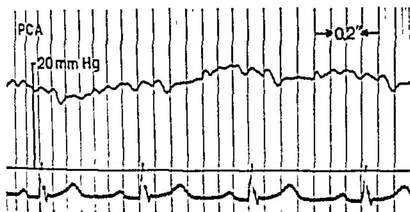


Fig. 724 —PCA pressure curve in primary pulmonary hypertension. Girl, aged 16 (B.A 57730) The curve does not have the typical appearance of an arterial pulse curve, but is damped (cf Fig 413, p 446).

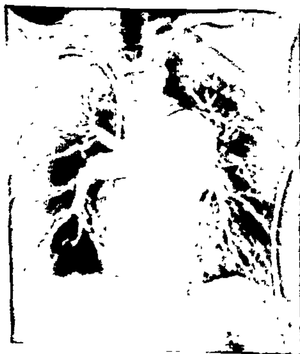


Fig. 725 —Primary pulmonary hypertension Boy, aged 11 (L.O 430514) Hypertrophy of right ventricle, considerable dilatation of main trunk of pulmonary artery and its main branches abrupt diminution in vessel caliber in periphery of the hilum peripheral pulmonary vessels extremely narrow

ing is an indication of constrictive vascular changes between the PCV and the PCA position.

Werkö and Eliasch (696) made exercise tolerance tests in combination with cardiac catheterization in cases of primary pulmonary hypertension. They found a rise in pressure in the pulmonary artery, but an inappreciable increase in the cardiac output and unchanged cardiopulmonary blood volume, which indicated vasoconstriction. On blocking the stellate ganglion, they found the pulmonary resistance to be somewhat increased. Hexamethonium bromide was given for some time, but it had no effect on the symptoms. Dresdale *et al.* (209) found that both Priscoline and ganglionic block (with TEB) caused a fall in the pulmonary artery pressure. The effect was, however, brief and did not appear with oral administration.

ANGIOCARDIOGRAPHY

Angiocardiographic examination with injection of the contrast medium into the right ventricle was performed in one of the cases. The ventricle was hypertrophied and showed increased trabeculation. The ventricular septum was intact, and the presence of a septal defect can be regarded as ruled out. Had it been present, it should have been visualized under the existing pressure conditions in the right ventricle.

There was definite dilatation of both the main trunk and the main branches of the pulmonary artery. Branching in the periphery of the hilum—corresponding to the transition into branches of the third order—was associated with an abrupt diminution in the caliber of the vessels (Fig. 725). The narrow peripheral vessels of the lungs had an otherwise normal appearance, and their outlines were regular.

31 "Idiopathic" Dilatation of the Pulmonary Artery

IN THE presence of a left to right shunt, the main trunk of the pulmonary artery is dilated because of an abnormally large flow, and in pulmonary stenosis, to a great extent because of turbulence. Even in completely healthy individuals, the main trunk of this artery may be found to be strikingly wide. It should then be regarded as an extreme normal variant The more thoroughly the patients are examined, the smaller will be the number of cases classified as "idiopathic" dilatation of the pulmonary artery. It is especially difficult to rule out mild pulmonary stenosis (282, 662) A pressure gradient of 10 to 15 mm Hg between the pulmonary artery and the right ventricle has often been noted (299, 373) A gradient of this order of magnitude under basic conditions should be considered pathologic. Normally, the rate of flow is not sufficiently high to produce a pressure of velocity of this order of magnitude (364).

Our series contains five cases in which the pulmonary artery was conspicuously wide although no other abnormal feature could be demonstrated

WOMAN, AGED 21 YEARS (E S 301018).—A

in the right ventricle (17/2 mm Hg) and pulmonary artery (15/2 mm Hg).

GIRL, AGED 15 YEARS (G G 380213).—After scarlet fever at the age of 7 years, she had contracted acute nephritis; since then she had frequently had proteinuria in association with throat infections She presented a number of



Fig. 726.—"Idiopathic" dilatation of the pulmonary artery. Girl, aged 6 (M V. 470521). Appearance of the heart is normal, apart from a distinct bulge in the border at the site of the main trunk of the pulmonary artery. Normal vascularity of lungs

nervous symptoms and often complained of breathlessness on exertion She was sent for a cardiologic examination because a wide "pulmonary arc" had been found on the roentgenogram The cardiac findings were normal, as was the *electrocardiogram* *Cardiac catheter-*

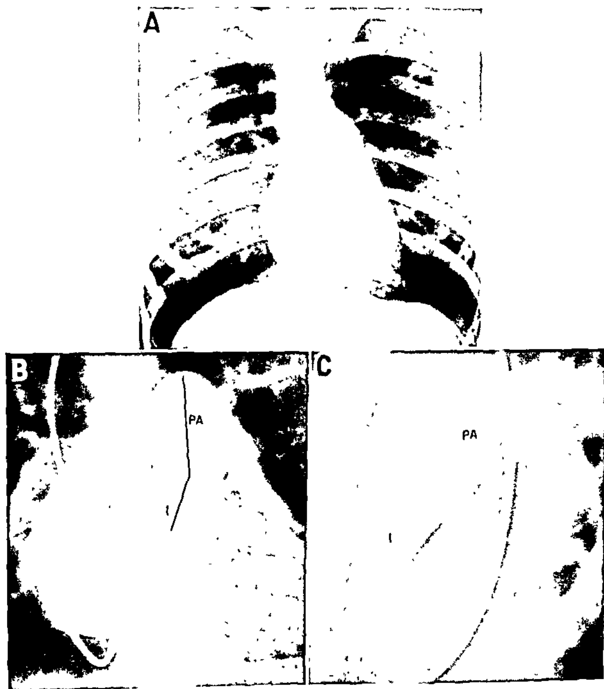


Fig. 727.—"Idiopathic" dilatation of the pulmonary artery Girl, aged 15 (G G 380213). Bulge in the superior part of the left cardiac border is due wholly to the main trunk of the pulmonary artery Its width lies within the normal range of variations The main trunk possibly ascends somewhat farther to the left than usual Otherwise, appearance of the heart is normal, as is vascularity of the lungs *I*, infundibulum, *PA*, pulmonary artery

ization disclosed no shunts, pressure in the right ventricle was 20/3 mm Hg and in the pulmonary artery 23/6 mm Hg The blood pressure was 110/70 mm Hg in the arm and 145/80 mm Hg in the leg Nothing abnormal was found in the urine.

Boy, aged 18 years (S.B 340814) —A murmur was detected when he was 5 years old, but he had never had cardiac symptoms or signs and was able to participate in strenuous sports Examination revealed a short, faint systolic murmur; it was regarded as physiologic. A loud, early systolic sound was heard over the pulmonary area (417), and the sec-

ond sound was split. The *electrocardiogram* was normal *Cardiac catheterization* revealed no shunts Pressure in the right ventricle was 22/2 mm Hg and in the pulmonary artery 17/4.

GIRL, AGED 6 YEARS (M.V. 470521).—On roentgenologic examination of the lungs made because of a respiratory tract infection, a wide pulmonary artery was found. She had never had cardiac symptoms or signs. The physical [illegible]

[illegible] 9 mm Hg

GIRL, AGED 14 YEARS (R-M.B. 410309).—A murmur was heard on routine examination when she was 10 years old She had never had any cardiac symptoms Examination disclosed a very faint early-systolic murmur and an early systolic sound over the pulmonary area The [illegible]

ROENTGENOLOGIC EXAMINATION

Particularly in childhood, a prominent pulmonary artery is a common isolated roentgenologic finding. It may be due to the position of the heart and need not imply that the artery is dilated In a classification of "idiopathic" pulmonary dilatation as a special entity, the criteria are essentially roentgenologic and are based on estimation of the pulmonary artery as dilated. The normal range of variations in its width are unknown; consequently, the estimation is made on somewhat arbitrary grounds.

All of our cases exhibited a pulmonary artery that was wider than the average (Figs 726 and 727), whereas the central and peripheral pulmonary vessels had the normal appearance. The pulsations over the pulmonary artery were not enlarged, a feature that has been stated by other authors to be typical (373). In the two cases in which electrokymography was performed, the tracings had a normal appearance. In none of the cases were the configuration and size of the heart abnormal.

ANGIOCARDIOGRAPHY

Angiocardiographic examination by injection of the contrast medium into the right ventricle in two of the cases revealed no abnormal conditions The shape of the pulmonary artery was normal (Fig. 727), and its increased width was regarded as a normal variation.

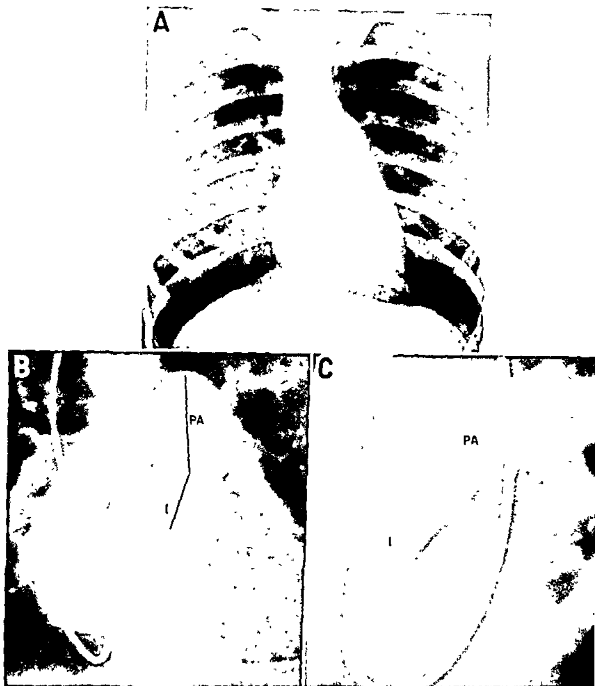


Fig. 727.—"Idiopathic" dilatation of the pulmonary artery Girl, aged 15 (G G 380213 Bulge in the superior part of the left cardiac border is due wholly to the main trunk of the pu monary artery Its width lies within the normal range of variations The main trunk possibly a cends somewhat farther to the left than usual Otherwise, appearance of the heart is normal, as vascularity of the lungs *I*, infundibulum, *PA*, pulmonary artery

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